Spevigo® (spesolimab)
(Intravenous/Subcutaneous)

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I. Length of Authorization [1]

- Treatment of GPP Flare: Coverage will be provided for two doses (900mg each) and may not be renewed.
- Treatment of GPP When Not Experiencing a Flare: Coverage will be provided for 6 months and may be renewed.

II. Dosing Limits

A. Quantity Limit (max daily dose) [NDC Unit]:

- Spevigo 150 mg/mL single-dose pre-filled syringe for subcutaneous use
  - Loading: 4 syringes x 1 dose only
  - Maintenance: 2 syringes every 4 weeks
- Spevigo 450 mg/7.5 mL single-dose vial for intravenous use: 4 vials one time only

B. Max Units (per dose and over time) [HCPCS Unit]:

- Treatment of GPP Flare *(IV formulation ONLY)*
  - 900 billable units (900 mg) on day 1 and 8
- Treatment of GPP When Not Experiencing a Flare *(SQ formulation ONLY)*
  - Loading: 600 mg x 1 dose only
  - Maintenance: 300 mg every 28 days

III. Initial Approval Criteria [1]

Coverage is provided in the following conditions:

- Patient is at least 12 years of age and weighs at least 40 kg; **AND**
- Patient has received all age-appropriate vaccinations according to current immunization guidelines prior to initiating treatment; **AND**

Universal Criteria [1-3,6]

- Patient does not have any of the following conditions:
  - Synovitis-acne-pustulosis-hyperostosis-osteitis (SAPHO) syndrome
  - Primary erythrodermic psoriasis vulgaris
  - Primary plaque psoriasis vulgaris without presence of pustules or with pustules that are restricted to psoriatic plaques**
  - Drug-triggered Acute Generalized Exanthematous Pustulosis (AGEP)**; **AND**
- Patient has been evaluated and screened for the presence of latent tuberculosis (TB) infection prior to initiating treatment and will receive ongoing monitoring for presence of TB during treatment; **AND**
- Patient does not have an active infection, including clinically important localized infections; **AND**
- Patient will not receive live vaccines (viral and/or bacterial) during therapy; **AND**
- Patient is not on concurrent treatment with an IL-inhibitor, TNF-inhibitor, biologic response modifier or other non-biologic agent (e.g., apremilast, abrocitinib, tofacitinib, baricitinib, upadacitinib, deucravacitinib, etc.); **AND**
- Patient will not be on concomitant treatment with systemic immunosuppressants (e.g., retinoids, cyclosporine, methotrexate, etc.) or other topical agents (e.g., corticosteroids, calcipotriene, tacrolimus, etc.); **AND**

***NOTE:** Only applies to patients receiving treatment for a GPP flare*

Generalized Pustular Psoriasis (GPP) † Φ [1-3,6]

- Patient is experiencing an acute, moderate-to-severe intensity disease flare; **AND**
  - Presence of a disease flare is confirmed by the following:
    - Generalized Pustular Psoriasis Physician Global Assessment (GPPPGA) total score of at least 3 (moderate) [the total GPPPGA score ranges from 0 (clear) to 4 (severe)] ¥; **AND**
    - Presence of fresh pustules (new appearance or worsening of pustules); **AND**
    - GPPPGA pustulation sub score of at least 2 (mild); **AND**
    - At least 5% of body surface area (BSA) covered with erythema and the presence of pustules; **OR**
- Patient is NOT currently experiencing a disease flare; **AND**

- Physician has assessed baseline disease severity utilizing an objective measure/tool (e.g., GPPPGA, Dermatology Quality of Life Index (DLQI), Psoriasis Symptom Scale, etc.); **AND**
- Patient has a GPPPGA total score of 0 or 1 **¥**; **AND**
- Patient has least 2 GPP flares of moderate-to-severe intensity with fresh pustulation (new appearance or worsening) in the past

¥ Physician's Global Assessment for Generalized Pustular Psoriasis (GPPPGA) [7]
Erythema 0 = Clear: Normal or post-inflammatory hyperpigmentation 1 = Almost Clear: Faint, diffuse pink or slight red 2 = Mild: Light red 3 = Moderate: Bright red 4 = Severe: Deep fiery red
Pustules 0 = Clear: No visible pustules 1 = Almost Clear: Low density occasional small discrete (non-coalescent) pustules 2 = Mild: Moderate density grouped discrete small pustules (non-coalescent) 3 = Moderate: High density pustules with some coalescence 4 = Severe: Very high-density pustules with pustular lakes
Scaling/crusting 0 = Clear: No scaling and no crusting 1 = Almost Clear: Superficial focal scaling or crusting restricted to periphery of lesions 2 = Mild: Predominantly fine scaling or crusting 3 = Moderate: Moderate scaling or crusting covering most or all of lesions 4 = Severe: Severe scaling or crusting covering most or all lesions
*Composite mean score = (erythema + pustules + scaling)/3 Total GPPGA score given is: 0 if mean is 0 for all three components, 1 if mean is 0 to <1.5, 2 if mean is 1.5 to <2.5, 3 if mean is 2.5 to <3.5, 4 if mean is ≥3.5

† FDA Approved Indication(s); **‡** Compendia Recommended Indication(s); **Φ** Orphan Drug

IV. Renewal Criteria [1]

Coverage can be renewed based upon the following criteria:

- Patients continues to meet universal and other indication-specific relevant criteria identified in section III; **AND**
- Absence of unacceptable toxicity from the drug. Examples of unacceptable toxicity include: infections, hypersensitivity reactions [including anaphylaxis and delayed reactions such as drug reaction with eosinophilia and systemic symptoms (DRESS)], etc.; **AND**

Treatment of GPP Flare

- Coverage may not be renewed

Treatment of GPP When Not Experiencing a Flare

- Disease response compared to baseline, as indicated by a decrease in number and/or frequency of GPP flares, stabilization or improvement in GPPPGA total score , improvement in Dermatology Quality of Life Index (DLQI), and/or improvement in Psoriasis Symptom Scale (PSS)

Initiating/Reinitiating Subcutaneous Maintenance Therapy after Treatment of a GPP Flare

- After receiving intravenous treatment for a GPP flare, patients may be initiated on subcutaneous maintenance therapy *(Refer to Section III for criteria and Section V for dosing)*; **OR**
- Patients experiencing a GPP flare while receiving subcutaneous maintenance therapy may receive up to two intravenous doses to treat the flare *(Refer to Section III for criteria and Section V for dosing)*

V. Dosage/Administration [1]

Indication	Dose
Generalized Pustular Psoriasis (GPP)	Treatment of GPP Flare ***(IV administration ONLY)*** • Administer as a single 900 mg dose by intravenous infusion over 90 minutes. • If GPP flare symptoms persist, an additional intravenous 900 mg dose may be administered one week after the initial dose. Treatment of GPP When Not Experiencing a Flare ***(SQ administration ONLY)*** • Administer a loading dose of 600 mg (four 150 mg injections) followed by 300 mg (two 150 mg injections) subcutaneously 4 weeks later and every 4 weeks thereafter. Initiating or Reinitiating **Subcutaneous** Spevigo After Treatment of a GPP Flare with **Intravenous** Spevigo • Four weeks after treatment of a GPP flare with **intravenous** Spevigo, initiate or reinitiate **subcutaneous** Spevigo for treatment of GPP at a dose of 300 mg (two 150 mg injections) administered every 4 weeks. • A subcutaneous loading dose is not required following treatment of a GPP flare with intravenous Spevigo.
NOTE: • Intravenous infusion of Spevigo is only to be administered by a healthcare professional in a healthcare setting. • The 600 mg subcutaneous loading dose of Spevigo is to be administered by a healthcare professional. For subsequent 300 mg doses, if the healthcare professional determines that it is appropriate, a patient 12 years of age and older may self-inject or the caregiver may administer Spevigo after proper training in subcutaneous injection technique. In pediatric patients 12 to 17 years of age, administer Spevigo under the supervision of an adult.	

VI. Billing Code/Availability Information

HCPCS Code(s):

- J1747 – Injection, spesolimab-sbzo, 1 mg; 1 billable unit = 1 mg (IV formulation ONLY)
- J3490 – Unclassified Drugs *(SQ formulation ONLY)*

NDC(s):

- Spevigo 150 mg/mL two-pack single-dose pre-filled syringe for subcutaneous use: 0597-0620-xx
- Spevigo 450 mg/7.5 mL (60 mg/mL) two-pack single-dose vial for intravenous use: 00597-0035-xx

VII. References

1. Spevigo [package insert]. Ridgefield, CT; Boehringer Ingelheim Pharmaceuticals, Inc.; March 2024. Accessed March 2024.
2. Bachelez H, Choon SE, Marrakchi S, et al; Effisayil 1 Trial Investigators. Trial of Spesolimab for Generalized Pustular Psoriasis. N Engl J Med. 2021 Dec 23;385(26):2431-2440. doi: 10.1056/NEJMoa2111563.
3. Choon SE, Lebwohl MG, Marrakchi S, et al. Study protocol of the global Effisayil 1 Phase II, multicentre, randomised, double-blind, placebo-controlled trial of spesolimab in patients with generalized pustular psoriasis presenting with an acute flare. BMJ Open. 2021 Mar 30;11(3):e043666. doi: 10.1136/bmjopen-2020-043666.
4. Navarini AA, Burden AD, Capon F, et al. European consensus statement on phenotypes of pustular psoriasis. J Eur Acad Dermatol Venereol. 2017 Nov;31(11):1792–1799. Crossref. PubMed. ISI.
5. Fujita H, Terui T, Hayama K, et al. Japanese guidelines for the management and treatment of generalized pustular psoriasis: the new pathogenesis and treatment of GPP. J Dermatol. 2018 Nov;45(11):1235–1270. Crossref. PubMed. ISI.
6. Morita A, Choon SE, Bachelez H, et al. Design of Effisayil™ 2: A Randomized, Double-Blind, Placebo-Controlled Study of Spesolimab in Preventing Flares in Patients with Generalized Pustular Psoriasis. Dermatol Ther (Heidelb). 2023 Jan;13(1):347-359. doi: 10.1007/s13555-022-00835-6. Epub 2022 Nov 5. PMID: 36333618; PMCID: PMC9823166.
7. Burden AD, Bachelez H, Choon SE, et al. The Generalized Pustular Psoriasis Physician Global Assessment (GPPGA) score: online assessment and validation study of a specific measure of GPP disease activity, British Journal of Dermatology, Volume 189, Issue 1, July 2023, Pages 138–140, https://doi.org/10.1093/bjd/ljad071.

Appendix 1 – Covered Diagnosis Codes

ICD-10	ICD-10 Description
L40.1	Generalized pustular psoriasis

Appendix 2 – Centers for Medicare and Medicaid Services (CMS)

The preceding information is intended for non-Medicare coverage determinations. Medicare coverage for outpatient (Part B) drugs is outlined in the Medicare Benefit Policy Manual (Pub. 100-2), Chapter 15, §50 Drugs and Biologicals. In addition, National Coverage Determinations (NCDs) and/or Local Coverage

Determinations (LCDs) may exist and compliance with these policies is required where applicable. Local Coverage Articles (LCAs) may also exist for claims payment purposes or to clarify benefit eligibility under Part B for drugs which may be self-administered. The following link may be used to search for NCD, LCD, or LCA documents: https://www.cms.gov/medicare-coverage-database/search.aspx. Additional indications, including any preceding information, may be applied at the discretion of the health plan.

Medicare Part B Covered Diagnosis Codes (applicable to existing NCD/LCD/LCA): N/A

Medicare Part B Administrative Contractor (MAC) Jurisdictions		
Jurisdiction	**Applicable State/US Territory**	**Contractor**
E (1)	CA, HI, NV, AS, GU, CNMI	Noridian Healthcare Solutions, LLC
F (2 & 3)	AK, WA, OR, ID, ND, SD, MT, WY, UT, AZ	Noridian Healthcare Solutions, LLC
5	KS, NE, IA, MO	Wisconsin Physicians Service Insurance Corp (WPS)
6	MN, WI, IL	National Government Services, Inc. (NGS)
H (4 & 7)	LA, AR, MS, TX, OK, CO, NM	Novitas Solutions, Inc.
8	MI, IN	Wisconsin Physicians Service Insurance Corp (WPS)
N (9)	FL, PR, VI	First Coast Service Options, Inc.
J (10)	TN, GA, AL	Palmetto GBA
M (11)	NC, SC, WV, VA (excluding below)	Palmetto GBA
L (12)	DE, MD, PA, NJ, DC (includes Arlington & Fairfax counties and the city of Alexandria in VA)	Novitas Solutions, Inc.
K (13 & 14)	NY, CT, MA, RI, VT, ME, NH	National Government Services, Inc. (NGS)
15	KY, OH	CGS Administrators, LLC