Spevigo® (spesolimab)
(Intravenous/Subcutaneous)

Effective Date: 04/01/2023
Review Date: 03/16/2023, 12/07/2023, 01/04/2024, 07/17/2024
Pharmacy Scope for Subcutaneous (SC) and Intravenous (IV) Formulations: Medicaid

I. Length of Authorization

- Treatment of GPP Flare: Coverage will be provided for two IV doses (900mg each) for 1 month 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 (post limit of 4 ml per 28 days or daily dose of 0.143)
  - Maintenance: 2 syringes every 4 weeks (2 ml per 28 days or daily dose of 0.08)
- Spevigo 450 mg/7.5 mL single-dose vial for intravenous use: 4 vials one time only (30 ml total)

III. Initial Approval Criteria[1,2,4,5,6]

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:

Neighborhood Health Plan of Rhode Island ©2024
Proprietary & Confidential – Not for Distribution

- 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 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**
- Patient is not on concurrent treatment with a TNF-inhibitor, biologic response modifier or other non-biologic agent (i.e., Otezla (apremilast), Xeljanz (tofacitinib), Olumiant (baricitinib), Rinvoq (upadacitinib), 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 as defined by the following:
  - Documentation that patient has a known documented history of GPP (either relapsing [greater than 1 episode] or persistent [greater than 3 months]); **AND**
  - Documentation that patient is presenting with primary, sterile, macroscopically visible pustules (new or worsening) on non-acral skin (excluding cases where pustulation is restricted to psoriatic plaques); **AND**
  - Documentation that patient has at least one of the following documented:
    - IL36RN, CARD14, or AP1S3 gene mutation; **OR**
    - Skin biopsy confirming presence of Kogoj's spongiform pustules; **OR**
    - Systemic symptoms or laboratory abnormalities commonly associated with GPP flare (e.g., fever, asthenia, myalgia, elevated C-reactive protein [CRP], leukocytosis, neutrophilia [above ULN]); **OR**
    - GPP flare of moderate-to-severe intensity with at least 5% body surface area covered with erythema and the presence of pustules **AND** 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** GPPPGA pustulation sub score of at least 2 (mild); **OR**
- Patient is NOT currently experiencing a disease flare; **AND**
  - Documentation that patient has a known documented history of GPP (either relapsing [greater than 1 episode] or persistent [greater than 3 months]); **AND**
  - Documentation that 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**

Neighborhood Health Plan of Rhode Island ©2024
Proprietary & Confidential – Not for Distribution

- Documentation that patient has a GPPPGA total score of 0 (clear) or 1 (almost clear) **¥**; **AND**
- Documentation that patient meets either of the following:
    - Patient has a history of at least 2 GPP flares of moderate-to-severe intensity (e.g., at least 5% body surface area covered with erythema and the presence of pustules, Generalized Pustular Psoriasis Physician Global Assessment [GPPPGA] total score of at least 3 (moderate) **¥** and GPPPGA pustulation sub score of at least 2 (mild); **OR**
    - Patient has a history of flaring while on concomitant treatment (e.g., retinoids, methotrexate, cyclosporine).

¥ 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[1,2,4,5,6]

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:

Neighborhood Health Plan of Rhode Island ©2024
Proprietary & Confidential – Not for Distribution

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

- Documentation of 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[1,4]

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 ***(SC 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

Neighborhood Health Plan of Rhode Island ©2024
Proprietary & Confidential – Not for Distribution

	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:

- J1747 injection, spesolimab-sbzo, 1mg

NDC:

- 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 (SDV): 00597-0035-xx

VII. References

1. Spevigo [package insert]. Ridgefield, NJ; Boehringer Ingelheim Pharmaceuticals, Inc.; March 2024. Accessed July 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

Neighborhood Health Plan of Rhode Island ©2024
Proprietary & Confidential – Not for Distribution

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

Neighborhood Health Plan of Rhode Island ©2024
Proprietary & Confidential – Not for Distribution