Blue Cross Blue Shield Blue Care Network of Michigan

Nonprofit corporations and independent licensees of the Blue Cross and Blue Shield Association

Medical benefit drug policies are a source for BCBSM and BCN medical policy information only. These documents are not to be used to determine benefits or reimbursement. Please reference the appropriate certificate or contract for benefit information. This policy may be updated and therefore subject to change.

Effective Date: 10/03/2024

Scenesse® (afamelanotide injectable implant)

HCPCS: J7352

Policy:

Requests must be supported by submission of chart notes and patient specific documentation.

A. Coverage of the requested drug is provided when all the following are met:
   a. FDA approved indication
   b. FDA approved age
   c. Documentation of elevated erythrocyte metal-free protoporphyrin greater than or equal to 85% of the total erythrocyte protoporphyrin as determined by an accredited laboratory
      OR
   d. Documentation of a total erythrocyte protoporphyrin 50 – 100 times the upper limit of normal of the laboratory reference range as determined by an accredited laboratory
      OR
   e. Documentation of two mutations in the ferrochelatase (FECH) gene as determined by an accredited laboratory
   f. Patient has documented symptoms of erythropoietic protoporphyria phototoxicity
   g. No personal history of melanoma or other cancerous or precancerous skin lesions

B. Quantity Limitations, Authorization Period and Renewal Criteria
   a. Quantity Limits: Align with FDA recommended dosing
   b. Authorization Period: 6 months at a time
   c. Renewal Criteria: Documentation of improvement in the duration of sun exposure without pain

***Note: Coverage and approval duration may differ for Medicare Part B members based on any applicable criteria outlined in Local Coverage Determinations (LCD) or National Coverage Determinations (NCD) as determined by Center for Medicare and Medicaid Services (CMS). See the CMS website at http://www.cms.hhs.gov/. Determination of coverage of Part B drugs is based on medically accepted indications which have supported citations included or approved for inclusion determined by CMS approved compendia.

Background Information:

- Scenesse is an alpha-melanocyte stimulating hormone analog indicated to increase pain free light exposure in adult patients with a history of phototoxic reactions from erythropoietic protoporphyria (EPP).

- EPP is an autosomal recessive, childhood-onset, rare porphyria with severe phototoxic manifestations. On sun exposure, patients experience prodromal symptoms, including tingling, burning, or itching, that serve as a warning signal, as continued exposure to sunlight leads to severe phototoxic pain and erythema and swelling of the exposed skin. The phototoxic pain is not responsive to treatment, including narcotic analgesics, and the severe pain may last for days. After experiencing the disabling phototoxic attacks, patients typically develop an ingrained fear of exposure to sunlight, leading to a conditioned behavior of sun avoidance that limits their daily activities and markedly impairs their quality of life.

- A diagnosis of EPP can be made in multiple ways if a patient is showing signs and symptoms of the disease. Biochemical testing is the most common method through which EPP is diagnosed via an elevated erythrocyte metal-free protoporphyrin greater than or equal to 85% of the total erythrocyte protoporpyrin level. Per Dr. Robert Desnick of Mount Sinai Hospital in New York City, an expert in EPP, diagnosis can also be made using the total erythrocyte protoporphyrin. He stated EPP will have a total erythrocyte protoporphyrin 50 to 100 times the upper limit of normal of the laboratory reference range. Finally, genetic testing can be done. Genetic testing will show two mutations of the ferrochelatase (FECH) gene. While there are over 200 known mutations of the FECH gene, 98% of EPP patients have an IVS3-48T>C variant. All biochemical and genetic testing should be performed in a laboratory that specializes in and is accredited in porphyria testing.

- The efficacy and safety of Scenesse was established in two Phase 3 studies. Patients in the United States and European Union were randomly assigned 1:1 to receive a subcutaneous implant of afamelanotide or placebo every 60 days for a six-month and a nine-month period, respectively. Patients in the US received 3 doses and those in the EU 5 doses. The study was designed to take place mostly during the summer months. The primary efficacy endpoint was duration of direct sunlight exposure between 10:00 am and 3:00 pm in the EU trial and 10:00 am and 6:00 pm in US trial on days when no pain was experienced (pain score of 0). There were 167 patients enrolled, 74 patients in the EU trial and 93 patients in the U.S. trial. Patients were excluded if they had a history of melanoma or other cancerous or precancerous skin lesions. In the United States, the median duration of pain-free time was longer over a 6-month period in the afamelanotide group compared to placebo (64.1 hours vs. 40.5 hours). In the European Union, the median duration of pain-free survival was longer over a 9-month period in the afamelanotide group compared to placebo (6.0 vs. 0.75).

References:

1. Scenesse [prescribing information]. West Menlo Park, CA: Clinuvel, Inc.; October 2022.
2. Langendonk JG, Balwani M, Anderson KE, et al. Afamelanotide for erythropoietic protoporphyria. NEJM. 2015; 373: 48 – 59.
3. Balwani M. National Organization for Rare Diseases. Erythropoietic protoporphyria and x-linked protoporphyria. Available at: https://rarediseases.org/rare-diseases/erythropoietic-protoporphyria/. Accessed on: September 3, 2020.
4. Donker AE, Raymakers AEP, Valsveld LT, et al. Practice guidelines for the diagnosis and management of microcytic anemias due to genetic disorders of iron metabolism or heme synthesis. Blood. 2014 June 19; 123 (25): 3973 – 86.
5. Kushner JP and Phillips JD. Erythropoietic protoporphyria: multiple pathways to a common phenotype. Hematologist. 2013; 10 (5): 5 – 14.
6. Kopcke W and Krutmann J. Protection from sunburn with beta-carotene: a meta-analysis. Photochem Photobiol. 2008; 84 (2): 284.
7. Stozil U, Doss MO, and Schuppan D. Clinical guide and update on prophyrias. Gastroenter. 2019 August; 157 (2): 365 – 81.
8. Balwani M, Naik H, Anderson KE, et al. Clinical, biochemical, and genetic characterization of north american patients with erythropoietic protoporphyria and x-linked protoporphyria. JAMA Dermatol. 2017 Aug; 153 (8): 789 – 96.

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<tr><th colspan="3">Policy History</th></tr>
<tr><th>#</th><th>Date</th><th>Change Description</th></tr>
<tr><td>1.9</td><td>Effective Date: 10/03/2024</td><td>Annual review – no changes to criteria at this time</td></tr>
<tr><td>1.8</td><td>Effective Date: 10/12/2023</td><td>Annual review – no changes to criteria at this time</td></tr>
<tr><td>1.7</td><td>Effective Date: 10/06/2022</td><td>Annual review – no changes to criteria at this time</td></tr>
<tr><td>1.6</td><td>Effective Date: 10/07/2021</td><td>Annual review of criteria was performed, no changes were made.</td></tr>
<tr><td>1.5</td><td>Effective Date: 10/08/2020</td><td>Updated testing requirements to allow genetic testing and use of total protoporphyrin and also added expert opinion</td></tr>
<tr><td>1.4</td><td>Effective Date: 03/16/2020</td><td>UM medical management system update for MAPPO and BCNA
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<tr><td>1.3</td><td>Effective Date: 1/1/2020</td><td>UM medical management system update for BCBS
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<tr><td>1.2</td><td>Effective Date: 12/05/2019</td><td>Full drug review</td></tr>
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** The prescribing information for a drug is subject to change. To ensure you are reading the most current information it is advised that you reference the most updated prescribing information by visiting the drug or manufacturer website or http://dailymed.nlm.nih.gov/dailymed/index.cfm.*

Blue Cross Blue Shield/Blue Care Network of Michigan
Medication Authorization Request Form
Scenesse® (afamelanotide injectable implant) HCPCS CODE: J7352

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This form is to be used by participating physicians to obtain coverage for Scenesse. For commercial members only, please complete this form and submit via fax to 1-877-325-5979. If you have any questions regarding this process, please contact BCBSM Provider Relations and Servicing or the Medical Drug Helpdesk at 1-800-437-3803 for assistance.

PATIENT INFORMATION	PHYSICIAN INFORMATION
Name	**Name**
ID Number	**Specialty**
D.O.B. ☐Male ☐Female **Pt weight (in kg): _____** **Date recorded:_____**	**Address**
Diagnosis	**City /State/Zip**
Drug Name ☐	**Phone/Fax: P**: ( ) - **F**: ( ) -
Dose and Quantity	**NPI**
Directions	**Contact Person**
Date of Service(s)	**Contact Person Phone / Ext.**

STEP 1: DISEASE STATE INFORMATION

1. Is this request for: ☐ Initiation ☐ Continuation *Date patient started therapy:* _____

2. Please provide the NPI number for the place of administration: _____

3. **Initiation AND Continuation of therapy:**
   a. Please check the patient's diagnosis, ***(Please attach any tests confirming diagnosis)***:
      ☐ Increase pain free light exposure in adult patients with a history of phototoxic reactions from erythropoietic protoporphyria
      ☐ Other: _____
   a. Does the patient have documentation of elevated erythrocyte metal-free protoporphyrin greater than or equal to 85% of the total erythrocyte protoporphyrin as determined by an accredited laboratory?
      ☐ Yes ☐ No Comment:_____ Lab results: _____ Reference Range: _____ Date: _____
   b. Does the patient have documentation of a total erythrocyte protoporphyrin 50 – 100 times the upper limit of normal of the laboratory reference range as determined by an accredited laboratory?
      ☐ Yes ☐ No Comment:_____ Lab results: _____ Reference Range: _____ Date: _____
   b. Does the patient have documentation of two mutations in the ferrochelatase (FECH) gene as determined by an accredited laboratory?
      ☐ Yes ☐ No Comment: _____
   c. Does the patient have documentation of symptoms of erythropoietic protoporphyria phototoxicity?
      ☐ Yes ☐ No Comment: _____
   d. Does the patient have history of melanoma or other cancerous or precancerous skin lesions?
      ☐ Yes ☐ No Comment: _____

4. **Continuation of therapy: Scenesse start date:** _____
   a. Has the patient had documented clinical response (improvement) in the duration of sun exposure without pain?
      ☐ Yes ☐ No Comment: _____

Please add any other supporting medical information necessary for our review

Coverage will not be provided if the prescribing physician's signature and date are not reflected on this document.

☐ Request for expedited review: I certify that applying the standard review time frame may seriously jeopardize the life or health of the member or the member's ability to regain maximum function

Physician's Name **Physician Signature** **Date**

Step 2: Checklist	☐ Form Completely Filled Out ☐ Attached Chart Notes	☐ Genetic mutation
Step 3: Submit	**By Fax: BCBSM Specialty Pharmacy Mailbox 1-877-325-5979**	**By Mail: BCBSM Specialty Pharmacy Program P.O. Box 312320, Detroit, MI 48231-2320**