MASSACHUSETTS

Blue Cross Blue Shield of Massachusetts is an Independent Licensee of the Blue Cross and Blue Shield Association

Medical Policy

Scenesse (afamelanotide) for Treatment of Erythropoietic Protoporphyria (EPP)

Table of Contents

Policy Number: 077

BCBSA Reference Number: N/A
NCD/LCD: N/A

Related Policies

None

Policy[1]

Commercial Members: Managed Care (HMO and POS), PPO, and Indemnity Medicare HMO Blue℠ and Medicare PPO Blue℠ Members

Scenesse (afamelanotide) may be **MEDICALLY NECESSARY** for the treatment of erythropoietic protoporphyria (EPP) in adults 18 and over when all of the following criteria are met:

- History of phototoxic reaction with sun exposure, AND
- No evidence of significant liver involvement, AND
- Biochemically confirmed diagnosis of protoporphyria (total protoporphyrin level >500ug/dl (generally 1000-5000ug/dl), AND
- The prescriber is a specialist in the area of the patient's diagnosis (eg., dermatology), or the prescriber has consulted with a specialist in the area of the patient's diagnosis.

Retreatment is evaluated on an annual basis and may be **MEDICALLY NECESSARY** when all of the following criteria are met:

- Increase in pain free time during light/sun exposure, AND
- Reduction in number of phototoxic reactions or decrease in severity of phototoxic reactions from pretreatment baseline.

Prior Authorization Information

Inpatient

- For services described in this policy, precertification/preauthorization **IS REQUIRED** for all products if the procedure is performed **inpatient**.

Outpatient

- For services described in this policy, see below for products where prior authorization **might be required** if the procedure is performed **outpatient**.

	Outpatient
Commercial Managed Care (HMO and POS)	Prior authorization is **required**.
Commercial PPO and Indemnity	Prior authorization is **required**.
Medicare HMO Blue℠	Prior authorization is **required**.
Medicare PPO Blue℠	Prior authorization is **required**.

Prior Authorization Request Form: Scenesse (afamelanotide) for the treatment of Erythropoietic Protoporphyria

This form must be completed and faxed to: Medical and Surgical: 888-973-0726; Medicare Advantage: 1-800-447-2994.

Click here for Elzonris Scenesse (afamelanotide) for the treatment of Erythropoietic Protoporphyria prior authorization form #160

CPT Codes / HCPCS Codes / ICD Codes

Inclusion or exclusion of a code does not constitute or imply member coverage or provider reimbursement. Please refer to the member's contract benefits in effect at the time of service to determine coverage or non-coverage as it applies to an individual member.

Providers should report all services using the most up-to-date industry-standard procedure, revenue, and diagnosis codes, including modifiers where applicable.

The following codes are included below for informational purposes only; this is not an all-inclusive list.

The above medical necessity criteria MUST be met for the following codes to be covered for Commercial Members: Managed Care (HMO and POS), PPO, Indemnity, Medicare HMO Blue and Medicare PPO Blue:

HCPCS Codes:

HCPCS codes:	Code Description
J7352	Afamelanotide implant, 1 mg

ICD-10 Procedure Codes

ICD-10-PCS procedure codes:	Code Description
0HHPXYZ	Insertion of Other Device into Skin, External Approach

Description

Erythropoietic protoporphyria (EPP) is a rare condition that results in the build up of protoporphyrin in the blood marrow, red blood cells, blood plasma, skin and liver. EPP is caused by a genetic mutation on the FECH gene, which is responsible for the normal production of heme. The build-up of protoporphyrin can cause extreme skin reactions to sunlight, including severe pain, burns and blisters, and edema. Protoporphyrin is excreted through the liver putting patients with EPP at risk of chronic liver disease and recurrent gallstones. In extreme cases, patients may experience rapid acute liver failure. EPP affects 75,000-200,000 people worldwide and is often diagnosed in infancy and early childhood after noticeable symptoms due to sun exposure. Genetic testing can provide confirmatory diagnosis.

Patients with EPP are recommended to reduce exposure to sunlight and avoid sun exposure whenever possible. In some patients, the use of tanning creams demonstrated increase tolerance to sun exposure

by increasing the amount of pigmentation in the skin. Protoporphyrin levels should be monitored annually to assess for damage to the liver. Patients with severe cases of EPP may require liver and bone marrow transplants if there is significant liver damage. There are currently no oral or topical medications available for the treatment of EPP.

In October of 2019, the FDA approved a subcutaneous injectable medication that increases the pigmentation of the skin and allows patients to have longer periods of sun exposure without reaction. Scenesse is indicated for patients with confirmatory genetic testing and into the torso and is administered every 2 months. For individuals with EPP without significant liver involvement, the evidence includes three randomized controlled trials evaluating the effect of Scenesse in reducing phototoxic reactions and pain. In all of the available studies, patients were administered with either Scenesse or a "vehicle." In the Scenesse groups, patients reported no pain after spending a median of 6 hours per day outside in direct sunlight. In the control groups, patients reported "no pain" over the median time of .75 hours per day.

Summary

Erythropoietic Protoporphyria is a rare, genetic mutation that causes an abnormal production of protoporphyria in the bone marrow, red blood cells, plasma, skin and liver. As a result, patients experience phototoxic reactions when exposed to the sun. While symptoms vary, patients can experience pain, reddening of skin, blisters and burns to exposed skin and edema. In severe cases, patients might experience disease of the liver. There are currently no oral or topical medications that are approved to treat EPP. The FDA has approved a subcutaneous implant, Scenesse (afamelanotide), that is injected every 2 months in adult patients without significant liver involvement.

The evidence includes three randomized controlled trials. Of the 167 patients included, 86 patients with EPP without significant liver involvement received Scenesse and were evaluated on the primary endpoint of number of hours spent outside. Patients documented degree of pain during sun exposure each day. Over the course of 270 days, patients in the treatment group reported significant improvement over the controlled group for number of hours spent outside without symptom reaction and with reduced pain. The evidence is sufficient to determine that the technology results in a meaningful improvement in the net health outcome.

Policy History

Date	Action
2/2021	New medically necessary and investigational indications described. Clarified coding information. Effective 2/1/2021.
1/2021	Clarified coding information
5/2020	New medical policy describing investigational indications. Effective 5/1/2020.

Information Pertaining to All Blue Cross Blue Shield Medical Policies

Click on any of the following terms to access the relevant information:
Medical Policy Terms of Use
Managed Care Guidelines
Indemnity/PPO Guidelines
Clinical Exception Process
Medical Technology Assessment Guidelines

References

1. Clinuvel Pharmaceuticals, Inc. Prescribing Label: SCENESSE (afamelotide) implant, for subcutaneous use. Initial U.S. Approval: 2019.
2. Scenesse. Clinicaltrial.gov. Accessed on 1/20/2020. Available at: https://clinicaltrials.gov/ct2/show/NCT01097044?cond=scenesse&draw=2&rank=7
3. Scenesse. Clinicaltrial.gov. Accessed on 1/20/2020. Available at: https://clinicaltrials.gov/ct2/show/NCT04053270?cond=scenesse&draw=2&rank=5

4. National Institute of Health. Genetic and Rare Disease Information Center. Erythropoietic Protoporphyria. Accessed 1/3/2020. https://rarediseases.info.nih.gov/diseases/4527/erythropoietic-protoporphyria

Endnotes

[1] Based on expert opinion