MASSACHUSETTS

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

Medical Policy

Elzonris (tagraxofusp-erzs) for the Treatment of Blastic Plasmacytoid Dendritic Cell Neoplasm

Table of Contents

Policy Number: 009

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

Related Policies

Elzonris (tagraxofusp-erzs) for the Treatment of Blastic Plasmacytoid Dendritic Cell Neoplasm (BPDCN) Prior Authorization Request Form, #928

Policy[1]

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

The use of Elzonris (tagraxofusp-erzs) for the treatment of Blastic Plasmacytoid Dendritic Cell Neoplasm (BPDCN) in patients 2 years and older may be considered **MEDICALLY NECESSARY** when **ALL** of the following criteria are met:

- Confirmed diagnosis of BPDCN; **AND**
- Must be prescribed by or in consultation with an oncologist or hematologist; **AND**
- The patient has an ECOG performance score of 0-2; **AND**
- Documentation of serum albumin is greater than or equal to 3.2 g/dL, prior to the first dose of initial treatment cycle; **AND**
- Initial treatment cycle **MUST** be administered in an inpatient setting and patient will be monitored for at least 24 hours after last infusion:
    - Subsequent treatment cycles may be administered in an appropriate outpatient setting.

Elzonris (tagraxofusp-erzs) is considered **INVESTIGATIONAL** for all other indications, including but not limited to acute myeloid leukemia (AML), chronic myelomonocytic leukemia (CMML), and myelofibrosis.

Continuation of Elzonris (tagraxofusp-erzs) will be approved annually at the FDA-approved doses if there are no unacceptable toxicities and there is documented evidence of efficacy such as lack of disease progression.

Additional doses beyond the FDA-approved number of doses will require a new Prior Authorization request and review.

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**.

Requesting Prior Authorization Using Authorization Manager
Providers will need to use Authorization Manager to submit initial authorization requests for services. Authorization Manager, available 24/7, is the quickest way to review authorization requirements, request authorizations, submit clinical documentation, check existing case status, and view/print the decision letter. For commercial members, the requests must meet medical policy guidelines.

To ensure the service request is processed accurately and quickly:

- Enter the facility's NPI or provider ID for where services are being performed.
- Enter the appropriate surgeon's NPI or provider ID as the servicing provider, *not* the billing group.

Authorization Manager Resources

- Refer to our Authorization Manager page for tips, guides, and video demonstrations.

Complete Prior Authorization Request Form for Elzonris (928) using Authorization Manager.

For out of network providers: Requests should still be faxed to 888-973-0726.

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
J9269	Injection, tagraxofusp-erzs, 10 microgram

ICD-10 Procedure Codes

ICD-10-PCS procedure codes:	Code Description
XW033Q5	Introduction of Tagraxofusp-erzs Antineoplastic into Peripheral Vein, Percutaneous Approach, New Technology Group 5
XW043Q5	Introduction of Tagraxofusp-erzs Antineoplastic into Central Vein, Percutaneous Approach, New Technology Group 5

Description

Blastic Plasmacytoid Dendritic Cell Neoplasm (BPDCN) is a hematologic cancer that is caused by a malignant spread of plasmacytoid dendritic cells. These cells produce a large group of proteins called type 1 interferon that are responsible for regulating the immune system. In BPDCN, the overproduction of these cells results in an inflammatory immune response causing overexpression of interleukin-3 receptors (CD123). The overexpression of CD123 results in malignant growths on the skin which can appear in various shapes, sizes and colors. The disease may also progress to internal organs such as the lymph nodes, spleen, and central nervous system.

BPDCN is an aggressive cancer currently categorized as a subtype of high-risk acute myeloid leukemia. BPDCN is a rare condition representing less than 1% of acute myeloid cases and is most often diagnosed in males between 50-60 years old. The exact cause and prevalence of the disease is not yet understood. Current treatment options include intensive chemotherapy and hematopoietic stem cell transplantation. Clinical response rates and tolerability for current standards of care varies between patients. BPDCN is a fatal disease with a median survival rate of 8-14 months.

Elzonris is the first and only drug approved by the FDA for the treatment of BPDCN and has been evaluated in patients 2 years and older with relapsed/refractory and treatment naïve disease. Elzonris targets the CD123 receptors causing cell death by prohibiting protein synthesis. Common adverse events include hypoalbuminemia and capillary leak syndrome which can be fatal. Patients are infused with Elzonris on days 1-5 of a 21-day treatment cycle and can receive multiple treatment cycles each year.

Summary

For individuals with BPDCN, the evidence includes a 4-stage, non-randomized, multicenter study evaluating complete response rates and duration of response after treatment with Elzonris. The study includes both treatment naïve or refractory/relapsed patients. Complete response rates were measured by the absence of skin lesions and lack of lesions for areas where systemic/multisystemic disease was initially observed, and clinical complete response is defined as evidence of skin abnormality not associated with prior skin lesions resulting from active disease.

Tagraxofusp, a CD123 targeted therapy was evaluated in 89 patients with a median duration follow up of 34 months. The median overall survival was 15.8 months and overall response rate was 75%. 51% of patients who received a complete response were bridged to stem cell transplants. 70% of patients with baseline bone marrow disease achieved clearance of malignant disease. For relapsed refractory patients the overall response rate was 58% with a median overall survival of 8.2 months.

Policy History

Date	Action
8/2024	Annual policy review. Literature reviewed through August of 2024. No new references available. No changes to policy statements. 8/1/2024.
9/2023	Policy clarified to include prior authorization requests using Authorization Manager.
8/2023	Annual policy review. Literature reviewed through August 2023. No new references available. No changes to policy statements. 8/1/2023.
12/2021	Policy updated with literature review through December 2021. References updated. Policy statements unchanged. 12/1/2021.
11/2019	New medical policy describing medically necessary and investigational indications. Effective 11/1/2019.

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. Jen, E. Y., Gao, X., Li, L., et al. FDA approval Summary: Tagraxofusp-erza For Treatment of Blastic Plasmacytoid Dendritic Cell Neoplasm. Clinical Cancer Research. 26(3). February 2020. DOI: 10.1158/1078-0432.
2. Pemmaraju N, Konopleva, M. Approval of tagraxofusp-erzs for blastic plasmacytoid dendritic cell neoplasm. Blood Advances. 4(16) August 2020. https://doi.org/10.1182/bloodadvances.2019000173
3. Pemmaraju N, Lane A, Sweet K, et al. Tagraxofusp in Blastic Plasmacytoid Dendritic-Cell Neoplasm, New England Journal of Medicine (2019): 380:1628-1637. DOI: 10.1056/NEJMoa1815105
4. Stemline Therapeutics, Inc. Prescribing Label: ELZONRIS (tagraxofusp-erzs) injection, for intravenous use Initial U.S. Approval: 2018. https://www.accessdata.fda.gov/drugsatfda_docs/label/2018/761116s000lbl.pdf. Accessed 12/21/2018
5. Blastic Plasmacytoid Dendritic Cell Neoplasm (BPDCN) Center, Dana Farber Cancer Institute. Retrieved from: https://www.dana-farber.org/bpdcn-center/
6. Pemmaraju N. Blastic plasmacytoid dendritic cell neoplasm. Clin Adv Hematol Oncol. 2016;14:220-222.
7. Elzonris. Clinicaltrial.gov. Accessed on 1/30/19. Available at: https://clinicaltrials.gov/ct2/show/NCT02113982

Endnotes

[1] Based on expert opinion