Blue Cross Blue Shield Blue Care Network of Michigan

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RETIRED

Effective Date: 02/04/2021

Elzonris™ (tagraxofusp-erzs)

FDA approval: 12/21/2018
HCPCS: J9269, C9049
Benefit: Medical

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. Prescribed by or in consultation with an oncologist
   b. FDA approved age
   c. Diagnosis of blastic plasmacytoid dendritic cell neoplasm confirmed on biopsy and with immunohistochemical testing showing positivity for CD4, CD56, CD123, TCL-1 and negative for myeloperoxidase and lysozyme
   d. ECOG performance score of 0 –2
   e. Serum albumin ≥ 3.2 g/dL, serum creatinine ≤ 1.5 mg/dL, bilirubin ≤ 1.5 mg/dL, and AST/ALT ≤ 2.5 times the upper limit of normal prior to starting therapy
   f. Must have a left ventricular ejection fraction ≥ 40% and no clinically significant cardiac abnormalities on EKG

B. Quantity Limitations, Authorization Period and Renewal Criteria
   a. Quantity Limit: FDA approved dosing
   b. Initial Authorization Period: 6 months
   c. Renewal Criteria: No evidence of disease progression or unacceptable toxicity
   d. Renewal Authorization Period: 1 year

***Note: Coverage 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.

Therapeutic considerations:

A. **FDA approved indication /Diagnosis**
   a. For the treatment of blastic plasmacytoid dendritic cell neoplasm (BPDCN) in adult and pediatric patients two years and older.

**Please refer to most recent prescribing information.*

B. **Background Information**
   a. Blastic plasmacytoid dendritic cell neoplasm (BPDCN) is a rare hematologic neoplasm arising from the precursors of plasmacytoid dendritic cells
   b. It commonly manifests as cutaneous lesions with or without bone marrow involvement and leukemic dissemination
   c. It may present with features similar to, and can be mistaken for, certain diseases including acute myeloid leukemia, non-Hodgkin's lymphoma, acute lymphocytic leukemia, myelodysplastic syndromes, and chronic myelomonocytic leukemia, as well as other malignancies with skin manifestations
   d. The exact incidence is unknown, difficult to estimate, and thought to be under reported, but experts believe that it represents 0.7% of primary cutaneous skin lymphomas
   e. The diagnosis of BPDCN is based on the immunophenotypic diagnostic triad of CD123, CD4, and CD56
   f. The median overall survival reported by most groups for adult patients is eight to 14 months, despite use of multiagent cytotoxicchemotherapy
   g. No specific treatment guidelines are available for BPDCN
   h. Most chemotherapy used have been regimens following treatments used in acute myeloid leukemia (AML), acute lymphocytic leukemia (ALL), or lymphomas
   i. The optimal treatment for BPDCN is unknown, due to a paucity of data regarding therapy
   j. Based on retrospective studies, the clinical course and response to therapy appear to differ between children and adults
   k. For children less than 18 years of age:
      i. For newly diagnosed BPDCN, induction chemotherapy with a regimen similar to that used for high-risk acute lymphoblastic leukemia (ALL) is recommended
      ii. For those who achieve a complete remission (CR) in first remission, observation rather than hematopoietic cell transplantation (HCT) is suggested
      iii. Allogeneic HCT is offered to patients who relapse and achieve a second remission
   l. For adults
      i. For newly diagnosed BPDCN, induction chemotherapy with a regimen similar to that used for ALL, including central nervous system (CNS) prophylaxis or treatment, is suggested
      ii. In those who achieve a CR in first remission, allogeneic HCT rather than observation is recommended, with plans for HCT in second remission

C. **Efficacy**

**Please refer to most recent prescribing information.*

D. **Medication Safety Considerations**

Black Box Warning: Yes

**Please refer to most recent prescribing information.*

F. **Dosing and administration**
   a. Dosing:
      i. Premedicate with a H1-histamine antagonist, acetaminophen, corticosteroid, and a H2-histamine antagonist prior to each dose of Elzonris
      ii. Dose 12 mcg/kg IV over 15 minutes once daily on days 1-5 of a 21 day cycle
      iii. The first cycle must be administered in an inpatient setting

**Please refer to most recent prescribing information.*

G. **How supplied**
   a. 1000 mcg in 1 mL in a single-dose vial

References:

1. Elzonris [package insert]. New York, NY: Stemline Therapeutics, Inc.; December 2018.
2. Elzonris (tagraxofusp-erzs). IPD Analytics. January 2019.
3. Clinicaltrials.gov. SL-401 in patients with blastic plasmacytoid dendritic cell neoplasm or acute myeloid leukemia. (NCT02113982) Available at: https://clinicaltrials.gov/ct2/show/NCT02113982. Accessed on January 2, 2019.
4. Cesarman-Maus G, Lome C, Espinosa KA, et al. Clinical and pathological features and differential diagnosis of blastic plasmacytoid dendritic cell neoplasm (BPDCN): a series of cases. Blood. 2016; 128(22): 5183.
5. Leukemia and Lymphoma Society. Facts about blastic plasmacytoid dendritic cell neoplasm (BPDCN). May 2018. Available at: http://www.lls.org/sites/default/files/National/USA/Pdf/Publications/FSHP2_BPDCN.pdf. Accessed on January 3, 2019.
6. Sangle NA, Schmidt RL, Patel JL, et al. Optimized immunohistochemical panel to differentiate myeloid sarcoma from blastic plasmacytoid dendritic cell neoplasm. Modern Pathology. 2014; 27: 1137-43.
7. Pagana L, Valentini CG, Grammatico S, & Pulsoni A. Blastic plasmacytoid dendritic cell neoplasm: diagnostic criteria and therapeutical approaches. Brit J Hematology. 2016; 174: 188 – 202.
8. Pemmaraju N & Konopleva M. Treating blastic plasmacytoid dendritic cell neoplasm. The Hematologist. Sept-Oct 2018; 15(5).

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<tr><th>#</th><th>Date</th><th>Change Description</th></tr>
<tr><td>1.2</td><td>Effective Date: 02/04/2021</td><td>Policy to be retired and drug to be added to the Medical Oncology Drug Class Policy</td></tr>
<tr><td>1.1</td><td>Effective Date: 02/06/2020</td><td>Annual Review</td></tr>
<tr><td>1.0</td><td>Effective Date: 02/14/2019</td><td>New Policy
<table><tr><th>Line of Business</th><th>PA Required (Yes/No)</th></tr><tr><td>BCBS</td><td>No</td></tr><tr><td>BCN</td><td>No</td></tr><tr><td>MAPPO</td><td>TBD</td></tr><tr><td>BCNA</td><td>TBD</td></tr></table></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.*