BlueCross BlueShield Arizona

An Independent Licensee of the Blue Cross Blue Shield Association

PHARMACY COVERAGE GUIDELINES
SECTION: DRUGS

ORIGINAL EFFECTIVE DATE: 5/19/2022
LAST REVIEW DATE:
LAST CRITERIA REVISION DATE:
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VIJOICE® (alpelisib) oral

Coverage for services, procedures, medical devices and drugs are dependent upon benefit eligibility as outlined in the member's specific benefit plan. This Pharmacy Coverage Guideline must be read in its entirety to determine coverage eligibility, if any.

This Pharmacy Coverage Guideline provides information related to coverage determinations only and does not imply that a service or treatment is clinically appropriate or inappropriate. The provider and the member are responsible for all decisions regarding the appropriateness of care. Providers should provide BCBSAZ complete medical rationale when requesting any exceptions to these guidelines.

The section identified as "Description" defines or describes a service, procedure, medical device or drug and is in no way intended as a statement of medical necessity and/or coverage.

The section identified as "Criteria" defines criteria to determine whether a service, procedure, medical device or drug is considered medically necessary or experimental or investigational.

State or federal mandates, e.g., FEP program, may dictate that any drug, device or biological product approved by the U.S. Food and Drug Administration (FDA) may not be considered experimental or investigational and thus the drug, device or biological product may be assessed only on the basis of medical necessity.

Pharmacy Coverage Guidelines are subject to change as new information becomes available.

For purposes of this Pharmacy Coverage Guideline, the terms "experimental" and "investigational" are considered to be interchangeable.

This Pharmacy Coverage Guideline does not apply to FEP or other states' Blues Plans.

Information about medications that require precertification is available at www.azblue.com/pharmacy.

Some large (100+) benefit plan groups may customize certain benefits, including adding or deleting precertification requirements.

All applicable benefit plan provisions apply, e.g., waiting periods, limitations, exclusions, waivers and benefit maximums.

Precertification for medication(s) or product(s) indicated in this guideline requires completion of the request form in its entirety with the chart notes as documentation. **All requested data must be provided.** Once completed the form must be signed by the prescribing provider and faxed back to BCBSAZ Pharmacy Management at (602) 864-3126 or emailed to Pharmacyprecert@azblue.com. **Incomplete forms or forms without the chart notes will be returned.**

VIJOICE® (alpelisib) oral

Criteria:

- **Criteria for initial therapy:** Vijoice (alpelisib) is considered ***medically necessary*** and will be approved when **ALL** of the following criteria are met:

  1. Prescriber is a physician specializing in the patient's diagnosis or is in consultation with a Pediatrician or Geneticist

  2. Individual is 2 years of age or older

  3. A confirmed diagnosis of severe manifestations of PIK3CA-Related Overgrowth Spectrum (PROS) who require systemic therapy and there is documented evidence of mutation in the *PIK3CA* gene with confirmed diagnosis of **ONE** of the following:
     a. Fibroadipose hyperplasia or overgrowth
     b. Hemihyperplasia multiple lipomatosis
     c. CLOVES (congenital lipomatous overgrowth, vascular malformations, epidermal nevi, and skeletal anomalies) syndrome
     d. Macrodactyly
     e. Fibroadipose infiltrating lipomatosis/facial infiltrative lipomatosis
     f. Macrocephaly-capillary malformation (M0CM, MCAP)
     g. Dysplastic megalencephaly
     h. Klippel-Trenaunay syndrome

  4. **ALL** of the following **baseline tests** have been completed before initiation of treatment with continued monitoring as clinically appropriate:
     a. Fasting plasma glucose
     b. Hemoglobin A1c
     c. Negative pregnancy test in a woman of childbearing potential

  5. Will not be combined with other alpelisib containing products

  6. Individual does not have a history of Stevens-Johnson syndrome, erythema multiforme, or toxic epidermal necrolysis

  7. Individual does not have type 1 diabetes mellitus or uncontrolled type 2 diabetes mellitus

  8. There are no significant interacting drugs

  Initial approval duration: 6 months

- **Criteria for continuation of coverage (renewal request):** Vijoice (alpelisib) is considered ***medically necessary*** and will be approved when **ALL** of the following criteria are met:

  1. Individual continues to be seen by a physician specializing in the patient's diagnosis or is in consultation with a Pediatrician or Geneticist

VIJOICE® (alpelisib) oral

2. Individual's condition has responded while on therapy
    a. Response is defined as **BOTH** of the following:
        i. There is at least a 20% reduction in the target lesion volume
        ii. There is no presence of at least a 20% increase in any target lesion, progression of non-target lesions or appearance of new lesions

3. Individual has been adherent with the medication

4. Individual has not developed any significant adverse drug effects that may exclude continued use
    a. Significant adverse effect such as:
        i. Severe hypersensitivity
        ii. Stevens-Johnson syndrome
        iii. Erythema multiforme
        iv. Toxic epidermal necrolysis
        v. Drug reaction with eosinophilia and systemic symptoms (DRESS)
        vi. Severe hyperglycemia, despite treatment
        vii. Hyperglycemic hyperosmolar non-ketotic syndrome
        viii. Ketoacidosis
        ix. Pneumonitis/Interstitial lung disease
        x. Severe diarrhea, despite anti-diarrheal agents
        xi. Pancreatitis

5. Dose is at least 50 mg daily

6. Will not be combined with other alpelisib containing products

7. Individual does not have a history of Stevens-Johnson syndrome, erythema multiforme, or toxic epidermal necrolysis

8. Individual does not have type 1 diabetes mellitus or uncontrolled type 2 diabetes mellitus

9. There are no significant interacting drugs

Renewal duration: 12 months

- Criteria for a request for non-FDA use or indication, treatment with dosing, frequency, or duration outside the FDA-approved dosing, frequency, and duration, refer to one of the following Pharmacy Coverage Guideline:

1. **Off-Label Use of Non-Cancer Medications**

2. **Off-Label Use of Cancer Medications**

VIJOICE® (alpelisib) oral

Description:

Vijoice (alpelisib) is a kinase inhibitor that is indicated for the treatment of adult and pediatric patients 2 years of age and older with severe manifestations of PIK3CA-Related Overgrowth Spectrum (PROS) who require systemic therapy. This indication is approved under accelerated approval based on response rate and duration of response. Continued approval for this indication may be contingent upon verification and description of clinical benefit in a confirmatory trial(s).

Activating mutations in *PIK3CA* have been found to induce a spectrum of overgrowths and malformations comprising a wide group of clinically recognizable disorders commonly known as PROS.

PROS includes a range of clinical findings in which the core features are congenital or early-childhood onset of segmental/focal overgrowth with or without cellular dysplasia. Prior to the identification of *PIK3CA* as the causative gene, PROS was separated into distinct clinical syndromes based on the tissues and/or organs involved.

PROS groups lesions with heterogeneous, segmental, overgrowth phenotypes, with or without vascular anomalies, and includes fibroadipose hyperplasia or overgrowth, hemihyperplasia multiple lipomatosis, CLOVES (congenital lipomatous overgrowth, vascular malformations, epidermal nevi, and skeletal anomalies) syndrome, macrodactyly, fibroadipose infiltrating lipomatosis/facial infiltrative lipomatosis, macrocephaly-capillary malformation (M0CM, MCAP), dysplastic megalencephaly, and Klippel-Trenaunay syndrome.

Resources:

Vijoice (alpelisib) product information, revised by Novartis Pharmaceuticals Corporation 04-2022. Available at DailyMed http://dailymed.nlm.nih.gov. Accessed May 17, 2022.

Frieden IL, Chu DH. Klippel-Trenaunay syndrome: Clinical manifestations, diagnosis, and management. In: UpToDate, Levy ML, Corona R (Eds), UpToDate, Waltham MA.: UpToDate Inc. Available at http://uptodate.com. Topic last updated May 10, 2022. Accessed May 17, 2022.

Mirzaa G, Graham JM, Keppler-Noreuil K. PIK3CA-Related Overgrowth Spectrum. GeneReviews® - NCBI Bookshelf (nih.gov). National Institute of Health. National Library of Medicine. National Center for Biotechnology Information. Available at www.ncbi.nlm.nih.gov. Topic last updated December 23, 2021. Accessed May 17, 2022.