PHARMACY COVERAGE GUIDELINES
SECTION: DRUGS

ORIGINAL EFFECTIVE DATE: 5/19/2022
LAST REVIEW DATE:
LAST CRITERIA REVISION DATE:
ARCHIVE DATE:

CAMZYOS™ (mavacamten) 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.**

CAMZYOS™ (mavacamten) oral

Criteria:

- **Criteria for initial therapy:** Camzyos (mavacamten) 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 Cardiologist

2. Individual is 18 years of age or older weighing at least 45 kilograms

3. A confirmed diagnosis of symptomatic New York Heart Association (NYHA) Class II–III obstructive hypertrophic cardiomyopathy (HCM) to improve functional capacity and symptoms

4. **ALL** of the following **baseline tests** have been completed before initiation of treatment with continued monitoring as clinically appropriate:
   a. Echocardiogram shows left ventricular ejection fraction (LVEF) is at least 55%
   b. Left ventricular outflow tract (LVOT) peak gradient is at least 50 mmHg at rest or with provocation
   c. Negative pregnancy test in a woman of childbearing potential

5. Verification that the patient is enrolled in the CAMZYOS REMS PROGRAM

6. Documented failure, contraindication per FDA label, intolerance, or not a candidate to **ALL** the following:
   a. Beta-blocker either bisoprolol, carvedilol, or metoprolol
   b. Calcium channel blocker either verapamil or diltiazem
   c. Combination of beta-blocker and calcium channel blocker

7. There are **NO** FDA-label contraindications, such as: [**Note**: this criterion is waived if individual, provider, and Pharmacy are enrolled in CAMZYOS REMS PROGRAM]
   a. Moderate to strong CYP2C19 inhibitors or strong CYP3A4 inhibitors
   b. Moderate to strong CYP2C19 inducers or moderate to strong CYP3A4 inducers

8. Individual does not have any of the following:
   a. Amyloidosis
   b. Fabry disease
   c. Noonan syndrome with left ventricular hypertrophy

Initial approval duration: 6 months

- **Criteria for continuation of coverage (renewal request):** Camzyos (mavacamten) 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 Cardiologist

2. Individual's condition has responded while on therapy
   a. Response is defined as **THREE** of the following:

i. A decrease in LVOT gradient of at least 30 mmHg with valsalva maneuvers
ii. Maintains LVEF of at least 50%
iii. Improvement in at least one NYHA class
iv. One or more improvement on NYHA class and at least a 1.5 mL/kg/min improvement in exercise capacity through mixed venous oxygen tension (pVO2)
v. No worsening in NYHA class and at least a 3 mL/kg/min improvement in exercise capacity through mixed venous oxygen tension (pVO2)

3. Individual has been adherent with the medication and is enrolled in the CAMZYOS REMS PROGRAM

4. Individual has not developed any contraindications or other significant adverse drug effects that may exclude continued use
    a. Contraindications as listed in the criteria for initial therapy section
    b. Significant adverse effect such as:
        i. LVEF is less than 50%
        ii. Experiencing heart failure symptoms or worsening clinical status

5. There are no significant interacting drugs

6. Individual has not had two LVEF of less than 50% determinations while on a dose of 2.5 mg daily

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**

Description:

Camzyos (mavacamten) indicated for the treatment of adults with symptomatic New York Heart Association (NYHA) class II-III obstructive hypertrophic cardiomyopathy (HCM) to improve functional capacity and symptoms.

Camzyos (mavacamten) is an allosteric and reversible inhibitor selective for cardiac myosin. Camzyos (mavacamten) modulates the number of myosin heads that can enter “on actin” (power-generating) states, thus reducing the probability of force-producing (systolic) and residual (diastolic) cross-bridge formation. Excess myosin actin cross-bridge formation and dysregulation of the super-relaxed state are mechanistic hallmarks of HCM. Camzyos (mavacamten) shifts the overall myosin population towards an energy-sparing, recruitable, super-relaxed state. In HCM patients, myosin inhibition with Camzyos (mavacamten) reduces dynamic LVOT obstruction and improves cardiac filling pressures.

CAMZYOS™ (mavacamten) oral

<u>Resources</u>:

Camzyos (mavacamten) product information, revised by Bristol Myers Squibb 05-2022. Available at DailyMed http://dailymed.nlm.nih.gov. Accessed May 09, 2022.

Maron MS. Hypertrophic cardiomyopathy: Clinical manifestations, diagnosis, and evaluation. In: UpToDate, McKenna WJ, Yeon SB (Eds), UpToDate, Waltham MA.: UpToDate Inc. Available at http://uptodate.com. Topic last updated August 21, 2020. Accessed May 09, 2022.

Maron MS. Hypertrophic cardiomyopathy: Medical therapy for heart failure. In: UpToDate, McKenna WJ, Yeon SB (Eds), UpToDate, Waltham MA.: UpToDate Inc. Available at http://uptodate.com. Topic last updated August 08, 2019. Accessed May 09, 2022.