Pharmacy Medical Necessity Guidelines:
Zokinvy™ (lonafarnib)

Effective: April 14, 2026

Prior Authorization Required	√	Type of Review – Care Management	
Not Covered		Type of Review – Clinical Review	√
Pharmacy (RX) or Medical (MED) Benefit	RX	Department to Review	RXUM
These pharmacy medical necessity guidelines apply to the following: ☒ Tufts Health RITogether – A Rhode Island Medicaid Plan		**Fax Numbers:** RXUM: 617.673.0939	

Note: This guideline does not apply to Medicare Members (includes dual eligible Members).

OVERVIEW

FOOD AND DRUG ADMINISTRATION-APPROVED INDICATIONS

Zokinvy (lonafarnib) is a farnesyltransferase inhibitor indicated in patients 12 months of age and older with a body surface area of 0.39 m^2 and above to reduce the risk of mortality in Hutchinson-Gilford Progeria Syndrome and for treatment of processing-deficient Progeroid Laminopathies with either heterozygous LMNA mutation with progerin-like protein accumulation or homozygous or compound heterozygous *ZMPSTE24* mutations

Zokinvy (lonafarnib) is not indicated for other Progeroid Syndromes or processing-proficient Progeroid Laminopathies. Based upon its mechanism of action, Zokinvy (lonafarnib) would not be expected to be effective in these populations.

COVERAGE GUIDELINES

The plan may authorize coverage of **Zokinvy (lonafarnib)** for Members when the following criteria are met:

Hutchinson-Gilford Progeria Syndrome

1. Documented diagnosis of Hutchinson-Gilford Progeria Syndrome

AND

2. The patient is at least 12 months of age or older

AND

3. Prescribed by or in consultation with a specialist in progeria, genetics, or metabolic disorders

AND

4. Documented body surface area of at least 0.39 m^2

Progeroid Laminopathies

1. Documented diagnosis of processing deficient Progeroid Laminopathies

AND

2. The patient is at least 12 months of age or older

AND

3. Prescribed by or in consultation with a specialist in progeria, genetics, or metabolic disorders

AND

4. Documented body surface area of at least 0.39 m^2

AND

5. Documentation of **one (1)** of the following:
    a. Heterozygous LMNA mutation with progerin-like protein accumulation
    b. Homozygous or compound heterozygous *ZMPSTE24* mutations

LIMITATIONS

None

CODES

None

REFERENCES

1. Introne WJ, Merideth MA. Hutchinson-Gilford progeria syndrome. In: Post TW, ed. UpToDate. Waltham, MA: UpToDate, Inc. Accessed 18 February 2020.
2. Zokinvy (lonafarnib) [package insert]. Palo Alto, CA: Eiger BioPharmaceuticals, Inc.; March 2024

APPROVAL HISTORY

September 13, 2022: Reviewed by Pharmacy & Therapeutics Committee
- June 13, 2023: No changes
- April 9, 2024: No changes
- March 11, 2025: No changes (eff 3/11/25)
- April 14, 2026: No changes.

BACKGROUND, PRODUCT AND DISCLAIMER INFORMATION

Pharmacy Medical Necessity Guidelines have been developed for determining coverage for plan benefits and are published to provide a better understanding of the basis upon which coverage decisions are made. The plan makes coverage decisions on a case-by-case basis considering the individual member's health care needs. Pharmacy Medical Necessity Guidelines are developed for selected therapeutic classes or drugs found to be safe, but proven to be effective in a limited, defined population of patients or clinical circumstances. They include concise clinical coverage criteria based on current literature review, consultation with practicing physicians in the service area who are medical experts in the particular field, FDA and other government agency policies, and standards adopted by national accreditation organizations. The plan revises and updates Pharmacy Medical Necessity Guidelines annually, or more frequently if new evidence becomes available that suggests needed revisions.

For self-insured plans, coverage may vary depending on the terms of the benefit document. If a discrepancy exists between a Pharmacy Medical Necessity Guideline and a self-insured Member's benefit document, the provisions of the benefit document will govern.

Treating providers are solely responsible for the medical advice and treatment of members. The use of this policy is not a guarantee of payment or a final prediction of how specific claim(s) will be adjudicated. Claims payment is subject to member eligibility and benefits on the date of service, coordination of benefits, referral/authorization and utilization management guidelines when applicable, and adherence to plan policies and procedures and claims editing logic.

Provider Services