UnitedHealthcare Pharmacy
Clinical Pharmacy Programs

Program Number	2026 P 1354-6
Program	Prior Authorization/Notification
Medication	Zokinvy™ (lonafarnib)
P&T Approval Date	3/2021, 3/2022, 3/2023, 3/2024, 3/2025, 3/2026
Effective Date	6/1/2026

1. Background:

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 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
- Homozygous or compound heterozygous ZMPSTE24 mutations

Limitations of Use:
Zokinvy is not indicated for other progeroid syndromes or processing-proficient progeroid laminopathies. Based upon its mechanism of action, Zokinvy would not be expected to be effective in these populations.

Coverage Information:
Members will be required to meet the criteria below for coverage.

Some states mandate benefit coverage for off-label use of medications for some diagnoses or under some circumstances. Some states also mandate usage of other Compendium references. Where such mandates apply, they supersede language in the benefit document or in the notification criteria.

2. Coverage Criteria[a]:

A. Hutchinson-Gilford Progeria Syndrome

1. **Zokinvy** will be approved based on of the following criterion:

   a. Diagnosis of Hutchinson-Gilford Progeria Syndrome

   Authorization will be issued for 12 months.

B. Progeroid Laminopathies

1. **Zokinvy** will be approved based on of the following criteria:

   a. Diagnosis of processing deficient progeroid laminopathy

   -AND-

b. Documentation of **one** of the following:

i. Heterozygous *LMNA* mutation with progerin-like protein accumulation

-OR-

ii. Homozygous or compound heterozygous *ZMPSTE24* mutations

Authorization will be issued for 12 months.

[a] State mandates may apply. Any federal regulatory requirements and the member specific benefit plan coverage may also impact coverage criteria. Other policies and utilization management programs may apply.

3. Additional Clinical Rules:

- Notwithstanding Coverage Criteria, UnitedHealthcare may approve initial and re-authorization based solely on previous claim/medication history, diagnosis codes (ICD-10) and/or claim logic. Use of automated approval and re-approval processes varies by program and/or therapeutic class.
- Supply limits may be in place.

4. References:

1. Zokinvy [package insert]. Solana Beach, CA: Sentynl Therapeutics, Inc.; February 2025.

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Change Control	
3/2021	New program.
3/2022	Annual review. Updated reference formatting.
3/2023	Annual review with no change to coverage criteria. Added state mandate footnote.
3/2024	Annual review with no change to coverage criteria.
3/2025	Annual review with no change to coverage criteria. Updated background and reference.
3/2026	Annual review with no change to coverage criteria. Updated background and reference.