

Policy Name:	Zokinvy (lonafarnib)	Policy #:	2740P
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Purpose of the Policy

The purpose of this policy is to define coverage criteria for Zokinvy (lonafarnib) for the treatment of Hutchinson-Gilford progeria syndrome and processing-deficient progeroid laminopathies.

Statement of the Policy

Health Alliance Medical Plans will approve the use of Zokinvy (lonafarnib) under the pharmacy benefit if the following criteria are met.

Criteria

1. Coverage Criteria

- 1.1 Documented diagnosis of one of the following:
 - Hutchinson-Gilford progeria syndrome
 - Processing-deficient progeroid laminopathies with either:
 - Heterozygous LMNA mutation with progerin-like protein accumulation
 - Homozygous or compound heterozygous ZMPSTE24 mutations
- 1.2 Member is 12 months or older
- 1.3 Member has a Body Surface Area (BSA) $\geq 0.39\text{m}^2$
- 1.4 Ordered by, or in consultation with a specialist in progeria, genetics, or metabolic disorders
- 1.5 Review of chart notes documenting diagnosis and confirming that patient has met all above requirements for treatment with Zokinvy by both a pharmacist and medical director

2. Approval Period

- 2.1 Initial: 12 months
- 2.2 Reapproval: 12 months with documentation of improvement and member has seen provider within the past 90 days of treatment reapproval

CPT Codes

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HCPCS Codes

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References

1. Zokinvy (lonafarnib) [prescribing information]. Palo Alto, CA: Eiger Biopharmaceuticals Inc; March 2024
2. Gordon LB, Kleinman ME, Massaro J, et al. Clinical trial of the protein farnesylation inhibitors lonafarnib, pravastatin, and zoledronic acid in children with Hutchinson-Gilford Progeria syndrome. *Circulation*. 2016;134(2):114-125
3. Gordon LB, Shappell H, Massaro J, et al. Association of Lonafarnib Treatment vs No Treatment With Mortality Rate in Patients With Hutchinson-Gilford Progeria Syndrome. *JAMA* 2018; 319:1687.

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Pharmacy Drug Policy & Procedure

DISCLAIMER

This Medical Policy has been developed as a guide for determining medical necessity. The process of medical necessity review also entails review of the most recent literature and physician review. Medical Policy is not intended to dictate to providers how to practice medicine. Providers are expected to exercise their medical judgment in providing the most appropriate care. Health Alliance encourages input from providers when developing and implementing medical policies. Benefit determinations are based on applicable contract language in the member's Policy/ Subscription Certificate/ Summary Plan Description. This Medical Policy does not guarantee coverage. There may be a delay between the revision of this policy and the posting on the web. Please contact the Health Alliance Customer Service Department at 1-800-851-3379 for verification of coverage.