

**Zokinvy (lonafarnib)**  
Effective 07/01/2021

<b>Plan</b>	<input checked="" type="checkbox"/> MassHealth <input checked="" type="checkbox"/> Commercial/Exchange	<b>Program Type</b>	<input checked="" type="checkbox"/> Prior Authorization <input checked="" type="checkbox"/> Quantity Limit <input type="checkbox"/> Step Therapy
<b>Benefit</b>	<input checked="" type="checkbox"/> Pharmacy Benefit <input type="checkbox"/> Medical Benefit (NLX)		
<b>Specialty Limitations</b>	This medication has been designated specialty and must be filled at a contracted specialty pharmacy.		
<b>Contact Information</b>	<b>Specialty Medications</b>		
	All Plans	Phone: 866-814-5506	Fax: 866-249-6155
	<b>Non-Specialty Medications</b>		
	MassHealth	Phone: 877-433-7643	Fax: 866-255-7569
	Commercial	Phone: 800-294-5979	Fax: 888-836-0730
	Exchange	Phone: 855-582-2022	Fax: 855-245-2134
	<b>Medical Specialty Medications (NLX)</b>		
	All Plans	Phone: 844-345-2803	Fax: 844-851-0882
<b>Exceptions</b>	N/A		

### Overview

Zokinvy (lonafarnib) is a farnesyltransferase inhibitor to prevent farnesylation and subsequent accumulation of progerin and progerin-like proteins in the inner nuclear membrane in patients  $\geq 12$  months of age to reduce the risk of the risk of mortality in HGPS. Zokinvy is also indicated for the treatment of processing-deficient progeroid laminopathies with either heterozygous LMNA mutation with progerin-like protein accumulation, or homozygous or compound heterozygous ZMPSTE24 mutations.

### Coverage Guidelines

Authorization may be granted for members new to AllWays Health Partners who are currently receiving treatment with Zokinvy, excluding when the product is obtained as samples or via manufacturer’s patient assistance programs.

#### OR

Authorization may be granted for members when ALL the following criteria are met, and documentation is provided:

#### Hutchinson-Gilford Progeria Syndrome

1. The member has a diagnosis of Hutchinson-Gilford Progeria Syndrome has been confirmed with genetic testing indicating the patient has LMNA mutation.
2. The member is 12 months of age or older
3. The member has a body surface area (BSA) of 0.39 m<sup>2</sup> or above

#### Processing Deficient Progeroid Laminopathy with Progerin-Like Protein Accumulation

1. The member has a diagnosis of Processing Deficient Progeroid Laminopathy has been confirmed with genetic testing indicating the patient has heterozygous LMNA mutation.
2. The member is 12 months of age or older



3. The member has a BSA of 0.39 m<sup>2</sup> or above

**Processing Deficient Progeroid Laminopathy without Progerin-Like Protein Accumulation**

1. The member has a diagnosis of Processing Deficient Progeroid Laminopathy has been confirmed with genetic testing indicating the patient has homozygous or compound heterozygous ZMPSTE24 mutations
2. The member is 12 months of age or older
3. The member has a BSA of 0.39 m<sup>2</sup> or above

**Continuation of Therapy**

Reauthorization may be granted for members when ALL initial authorization criteria are met.

**Limitations**

1. Initial approvals and reauthorizations will be for 12 months.
2. The following quantity limits apply:

Zokinvy 50mg and 75mg capsules	120 capsules per 30 days
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**References**

1. Zokinvy [package insert]. Palo Alto, CA: Eiger BioPharmaceuticals, Inc.; November 2020.
2. Progeria Research Foundation (PRF). The Progeria Handbook: A Guide for Families & Health Care Providers of Children with Progeria. Second Edition. PRF. [https://www.progeriaresearch.org/wp-content/uploads/2019/03/PRF\\_Handbook\\_2019\\_eFile.pdf](https://www.progeriaresearch.org/wp-content/uploads/2019/03/PRF_Handbook_2019_eFile.pdf). Accessed November 27, 2020.
3. Gordon LB, Brown WT, Collins FS. Hutchinson-Gilford Progeria Syndrome. GeneReviews. University of Washington, Seattle; 2019.
4. AHFS DI (Adult and Pediatric) [database online]. Hudson, OH: Lexi-Comp, Inc.; [http://online.lexi.com/lco/action/index/dataset/complete\\_ashp](http://online.lexi.com/lco/action/index/dataset/complete_ashp) [available with subscription]. Accessed December 2, 2020.
5. Micromedex Solutions [database online]. Ann Arbor, MI: Truven Health Analytics Inc. Updated periodically. [www.micromedexsolutions.com](http://www.micromedexsolutions.com) [available with subscription]. Accessed December 2, 2020.

**Review History**

05/19/2021 – Created and Reviewed at May P&T. Effective 07/01/2021.

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