Mass General Brigham
Health Plan

Zokinvy (lonafarnib)
Effective 07/01/2021

Plan	☐ MassHealth UPPL ☒Commercial/Exchange	**Program Type**	☐ Prior Authorization ☐ Quantity Limit ☒ Step Therapy
Benefit	☒ Pharmacy Benefit ☐ Medical Benefit		
Specialty Limitations	This medication has been designated specialty and must be filled at a contracted specialty pharmacy.		
Contact Information	**Medical Benefit** **Pharmacy Benefit**	Phone: 833-895-2611 Phone: 800-711-4555	Fax: 888-656-6671 Fax: 844-403-1029
Exceptions	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 ≥ 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 the plan within the past 90 days who are currently receiving treatment, 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^2 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^2 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^2 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

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. 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 [available with subscription]. Accessed December 2, 2020.
5. Micromedex Solutions [database online]. Ann Arbor, MI: Truven Health Analytics Inc. Updated periodically. 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.

09/21/2022 – Reviewed at Sept P&T; Separated Comm/Exch vs MH policies; no clinical updates.