ConnectiCare

Commercial/Healthcare Exchange PA Criteria

Effective: 6/9/2021

Prior Authorization: ZOKINVY™ (lonafarnib)

Products Affected: ZOKINVY™ (lonafarnib) capsules, for oral use

Medication Description:
Lonafarnib inhibits farnesyltransferase to prevent farnesylation and subsequent accumulation of progerin and progerin-like proteins in the inner nuclear membrane.

Covered Uses:

- Hutchinson-Gilford Progeria Syndrome (HGPS)
- Processing-deficient Progeroid Laminopathies

Exclusion Criteria:
ZOKINVY is contraindicated in patients taking:

- Strong or moderate CYP3A inhibitors or inducers
- Midazolam
- Lovastatin, simvastatin, or atorvastatin

ZOKINVY is not indicated for other Progeroid Syndromes or processing-proficient Progeroid Laminopathies.

Required Medical Information:

1. Diagnosis
2. Genetic testing

Age Restrictions: 12 months of age and older

Prescriber Restrictions: Medication is prescribed by or in consultation with a geneticist, specialist in metabolic disorders, or pediatric cardiologist

Coverage Duration: 1 year for initial and continuation of therapy

Other Criteria:

I. Initial Approval Criteria
A. Hutchinson-Gilford Progeria Syndrome: Approve if the individual meets the following criteria (must meet all):

1. Patient has diagnosis of Hutchinson-Gilford Progeria Syndrome (HGPS); AND
2. Patient is ≥ 12 months of age; AND
3. Patient has a body surface area of ≥ 0.39 m2; AND
4. Requested dose is appropriate for patient's BSA and does not exceed the recommended dose of 150 mg/m2 twice daily

B. Processing-deficient Progeroid Laminopathies: Approve for 1 year if the individual meets the following criteria (must meet all):

1. Patient has diagnosis of processing-deficient Progeroid Laminopathies; AND
2. Patient has either Heterozygous *LMNA* mutation with progerin-like protein accumulation OR Homozygous or compound heterozygous *ZMPSTE24* mutations, confirmed by genetic testing
3. Patient is ≥ 12 months of age; AND
4. Patient has a body surface area of ≥ 0.39 m2; AND
5. Requested dose is appropriate for patient's BSA and does not exceed the recommended dose of 150 mg/m2 twice daily

II. Continued Therapy

1. Member is responding positively to therapy, as determined by the prescriber; AND
2. Member has not experienced unacceptable toxicity from the drug (for example nephrotoxicity, Myelosuppression, Electrolyte abnormalities, Increased liver enzymes, impaired fertility, or fetal harm)

Approval duration: 12 months

References:

1.Zokinvy (lonafarnib) [package insert]. Palo Alto, CA. Eiger BioPharmaceuticals, Inc. Updated February 10, 202. Accessed April 28, 2021.

Policy Revision history

Rev #	Type of Change	Summary of Change	Sections Affected	Date
1	New policy	New policy	All	6/9/2021