PHARMACY POLICY STATEMENT
North Carolina Marketplace

DRUG NAME	Zokinvy (lonafarnib)

Zokinvy is an oral farnesyltransferase inhibitor initially approved by the FDA in 2020. It is used for the treatment of certain mutations in processing-deficient Progeroid Laminopathies and to reduce the risk of mortality in Hutchinson-Gilford Progeria Syndrome. These are rare and fatal diseases of premature aging. Cardiovascular complications are the primary cause of mortality. Zokinvy is the first FDA approved disease-modifying treatment for these patients. Farnesyltransferase inhibition prevents farnesylation and subsequent accumulation of aberrant progerin and progerin-like proteins in the inner nuclear membrane.

Zokinvy (lonafarnib) will be considered for coverage when the following criteria are met:

Hutchinson -Gilford Progeria Syndrome

For initial authorization:

1. Member is at least 12 months of age; AND
2. Member has a body surface area (BSA) of 0.39 m^2 or greater; AND
3. Medication must be prescribed by or in consultation with a pediatrician, geneticist, cardiologist, or metabolic specialist; AND
4. Member has a diagnosis of Hutchinson-Gilford Progeria Syndrome confirmed by a known causative variant mutation in the LMNA gene (documentation required); AND
5. Member is NOT taking any of the following contraindicated drugs/drug classes:
   a) Strong or moderate CYP3A4 inhibitors or inducers;
   b) Midazolam;
   c) Lovastatin, simvastatin, or atorvastatin.
6. Dosage allowed/Quantity limit: Start at 115 mg/m^2 twice daily. After 4 months, increase to 150 mg/m^2 twice daily. Round all total doses to nearest 25 mg increment.

If all the above requirements are met , the medication will be approved for 12 months .

For reauthorization :

1. Member is tolerating therapy and is taking an appropriate dose.

If all the above requirements are met, the medication will be approved for an additional 12 months .

3. Medication must be prescribed by or in consultation with a pediatrician, geneticist, cardiologist, or metabolic specialist; AND
4. Member has a diagnosis of processing-deficient progeroid laminopathies confirmed by a known causative variant mutation in the LMNA gene (documentation required) with either:
   a) Heterozygous *LMNA* mutation with progerin-like protein accumulation, or
   b) Homozygous or compound heterozygous *ZMPSTE24* mutations
5. Member is NOT taking any of the following contraindicated drugs/drug classes: