SENTARA HEALTH PLANS

PHARMACY PRIOR AUTHORIZATION/STEP-EDIT REQUEST*

Directions: **The prescribing physician must sign and clearly print name (preprinted stamps not valid) on this request**. All other information may be filled in by office staff; **fax to 1-800-750-9692**. No additional phone calls will be necessary if all information (including phone and fax #s) on this form is correct. **If the information provided is not complete, correct, or legible, the authorization process can be delayed.**

Drug Requested: Zokinvy™ (lonafarnib)

MEMBER & PRESCRIBER INFORMATION: Authorization may be delayed if incomplete.

Member Name: _____

Member Sentara #: _____ **Date of Birth:** _____

Prescriber Name: _____

Prescriber Signature: _____ **Date:** _____

Office Contact Name: _____

Phone Number: _____ **Fax Number:** _____

DEA OR NPI #: _____

DRUG INFORMATION: Authorization may be delayed if incomplete.

Drug Form/Strength: _____

Dosing Schedule: _____ **Length of Therapy**: _____

Diagnosis: _____ **ICD Code, if applicable:** _____

Weight: _____ **Date:** _____

Dose does not exceed one of the following:

a. New starts or treated for less than 4 months: 230 mg/m^2 per day; rounded to the nearest 25mg dose (see tables) for a total of 4 months
   Maintenance after 4 months: 300mg/m^2 per day; rounded to the nearest 25mg dose (see tables)

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Initial BSA-based dosage for the starting dose of 115 mg/m^2 twice daily for 4 months					
BSA (m2)	**Total Daily Dosage Rounded to the nearest 25mg**	**Morning Dosing; Number of Capsules**		**Evening Dosing; Number of Capsules**	
		Zokinvy 50mg	**Zokinvy 75mg**	**Zokinvy 50mg**	**Zokinvy 75mg**
0.39-0.48	100	1		1	
0.49-0.59	125		1	1	
0.6-0.7	150		1		1
0.71-0.81	175	2			1
0.82-0.92	200	2		2	
0.93-1	225	1	1	2	

Maintenance BSA-based dosage of 150mg/m^2 twice daily					
BSA (m2)	**Total Daily Dosage Rounded to the nearest 25mg**	**Morning Dosing; Number of Capsules**		**Evening Dosing; Number of Capsules**	
		Zokinvy 50mg	**Zokinvy 75mg**	**Zokinvy 50mg**	**Zokinvy 75mg**
0.39-0.45	125		1	1	
0.46-0.54	150		1		1
0.55-0.62	175	2			1
0.63-0.7	200	2		2	
0.71-0.79	225	1	1	2	
0.8-0.87	250	1	1	1	1
0.88-0.95	275		2	1	1
0.96-1	300		2		2

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CLINICAL CRITERIA: Check below all that apply. All criteria must be met for approval. To support each line checked, all documentation, including lab results, diagnostics, and/or chart notes, must be provided or request may be denied.

Provider Please Note: Zokinvy™ is not indicated for other progeroid syndromes or processing proficient progeroid laminopathies. Based upon its mechanism of action, Zokinvy™ is not expected to be effective in these populations.

Initial authorization: 12 months

- ❑ Requested medication is prescribed by or in consultation with a specialist in progeria, genetics and/or metabolic disorders

AND

- ❑ Member is 12 months of age or older

AND

- ❑ Member does **NOT** have other non-laminopathy progeroid syndromes or processing proficient progeroid laminopathies or laminopathies with no progeria features (mutation in the LMNA gene with no clinical characteristic features)

AND

- ❑ Member has a body surface area of at least 0.39 m^2

AND

- ❑ Member will avoid concomitant therapy with **ALL** of the following:
  - ❑ midazolam
  - ❑ HMG-CoA reductase inhibitors (e.g., lovastatin, simvastatin or atorvastatin)
  - ❑ Strong or moderate CYP3A inhibitors (e.g., fluconazole, itraconazole, etc.)
  - ❑ Strong or moderate CYP3A inducers (e.g., rifampin, carbamazepine, St. John's Wart, etc)
  - ❑ Strong or moderate CYP2C9 inhibitors (e.g., voriconazole, metronidazole, fluvastatin, sulfamethoxazole, etc.)

OR

  - ❑ If concomitant therapy is unavoidable, the member will be monitored closely for adverse reaction and/or dose modifications will be implemented

AND

- ❑ Member has at least one of the following clinical features suggestive of progeria:
  - ❑ Characteristic facial appearance (e.g., micrognathia, prominent eyes and circumoral cyanosis)
  - ❑ Sclerodermatous skin changes (e.g., taut, thickened, fibrotic, indurated or rippled)
  - ❑ Alopecia and/or prominent scalp veins
  - ❑ Decreased joint range of motion and joint contractures
  - ❑ X-Ray findings (e.g., distal clavicular and terminal phalangeal resorption, coxa valga; delated/incomplete primary tooth eruption)
  - ❑ Severe atherosclerosis and/or cardiac disease (e.g., heart failure, stroke)

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AND

- ❑ Member has a diagnosis of one of the following **(documentation required)**:
  - ❑ Hutchinson-Gilfold progeria syndrome (HGPS)
  - ❑ Patient has had a confirmatory mutational analysis with a G608G mutation in a lamin A gene [LMNA gene] (e.g., c.1824C>T)

OR

- ❑ Member has a diagnosis of processing-deficient Progeroid Laminopathies **AND** one of the following:
  - ❑ Heterozygous LMNA mutation with progerin-like protein accumulation (e.g., pathogenic variant in either the exon 11 splice junction or intron 11 of LMNA gene)
  - ❑ Homozygous or compound heterozygous ZMPSTE24 mutations

Reauthorization approval: 12 months. Check below all that apply. All criteria must be met for approval. To support each line checked, all documentation, including lab results, diagnostics, and/or chart notes, must be provided or request may be denied.

- ❑ Absence of unacceptable toxicity from the drug. Examples of unacceptable toxicity include; severe laboratory abnormalities, severe nephrotoxicity, severe retinal toxicity, etc.

AND

- ❑ Disease response as indicated by improvement or stabilization in member's signs and/or symptoms and/or disease status (e.g., no new or worsening heart failure, no stroke incidence, evidence of decreased carotid-femoral pulse wave velocity, evidence of decreased carotid artery wall echo density)

AND

- ❑ If request is for a dose increase, new dose does not exceed 300mg/m^2 per day, rounded to the nearest 25mg dose (see tables above)

Medication being provided by a Specialty Pharmacy - PropriumRx

Not all drugs may be covered under every Plan

If a drug is non-formulary on a Plan, documentation of medical necessity will be required.

*****Use of samples to initiate therapy does not meet step edit/ preauthorization criteria.*****

****Previous therapies will be verified through pharmacy paid claims or submitted chart notes.****