

AvMed

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 1-305-671-0200.** 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 AvMed #: _____ 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/m² per day; rounded to the nearest 25mg dose (see tables) for a total of 4 months
Maintenance after 4 months: 300mg/m² per day; rounded to the nearest 25mg dose (see tables)

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<u>Initial BSA-based dosage for the starting dose of 115 mg/m² twice daily for 4 months</u>					
<u>BSA (m²)</u>	<u>Total Daily Dosage Rounded to the nearest 25mg</u>	<u>Morning Dosing; Number of Capsules</u>		<u>Evening Dosing; Number of Capsules</u>	
		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	

<u>Maintenance BSA-based dosage of 150mg/m² twice daily</u>					
<u>BSA (m²)</u>	<u>Total Daily Dosage Rounded to the nearest 25mg</u>	<u>Morning Dosing; Number of Capsules</u>		<u>Evening Dosing; Number of Capsules</u>	
		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 m²

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; delayed/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-Gilford 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/m² 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.****