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.**

Nitisinone Products

Drug Requested: select **ONE** drug below

❑ **Harliku** (nitisinone)	❑ **nitisinone capsules** (generic Orfadin®)
❑ **Nityr™** (nitisinone)	❑ **Orfadin®** (nitisinone) **capsules/suspension**

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:** _____

NPI #: _____

DRUG INFORMATION: Authorization may be delayed if incomplete.

Drug Name/Form/Strength: _____

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

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

Weight (if applicable): _____ **Date weight obtained:** _____

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.

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❑ **DIAGNOSIS: Hereditary Tyrosinemia Type 1 (HT-1)**

❑ **For generic nitisinone, Nityr & Brand Orfadin requests only**

Initial Authorization: 6 months

- ❑ Member must have a diagnosis of hereditary tyrosinemia type 1 (HT-1)
- ❑ Member must be using the prescribed medication as an adjunct to dietary restriction of tyrosine and phenylalanine
- ❑ Member's current patient's plasma tyrosine level is maintained at <500 mcmol/L **(submit documentation)**
- ❑ A laboratory test documenting baseline urinary or plasma succinylacetone level must have been completed within the last 30 days
- ❑ Member had had a baseline ophthalmologic examination with a normal slit lamp examination
- ❑ A complete blood count was completed within the last 30 days and has been submitted with request
- ❑ **For Nityr and Brand Orfadin capsule requests:** Member must have trial and failure of generic nitisinone capsules **(chart notes documenting clinically significant adverse effects and submission of completed MedWatch form are required for documentation)**
- ❑ **For Orfadin suspension requests:** Provider must submit documentation to confirm the member is unable to swallow nitisinone capsules **(submit documentation of intolerance to capsules)**
- ❑ Member's current weight must be noted in submitted chart notes; Maximum approved dosage will be 2 mg/kg/day

Reauthorization: 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.

- ❑ All initial authorization criteria continues to be met
- ❑ Member must meet **ONE** of the following **(submit documentation)**:
  - ❑ Member's urinary succinylacetone concentration has decreased to less than 1 mmol/mol creatinine from baseline level
  - ❑ Member's plasma succinylacetone concentration has decreased to less than 0.1 micromol/L from baseline level
- ❑ A complete blood count was completed within the last 30 days and has been submitted with request
- ❑ Member's current weight must be noted in submitted chart notes; Maximum approved dosage will be 2 mg/kg/day

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❑ **DIAGNOSIS: Alkaptonuria (AKU)**

❑ **For Harliku, generic nitisinone, Nityr or Orfadin requests**

Initial Authorization: 6 months

- ❑ Member is 18 years of age or older
- ❑ Prescribed by or in consultation with an endocrinologist or a metabolic or genetic disease specialist
- ❑ Member has a diagnosis of alkaptonuria as confirmed by **ONE** of the following **(submit documentation)**:
  - ❑ Baseline urinary HGA excretion greater than 0.4 g/24 hours
  - ❑ HGD (homogentisate 1,2-dioxygenase) biallelic gene mutation (mutations in both copies of the HGD gene) as evidenced by genetic testing
- ❑ **For Nityr and Brand Orfadin capsule requests:** Member must have trial and failure of generic nitisinone capsules **(chart notes documenting clinically significant adverse effects and submission of completed MedWatch form are required for documentation)**
- ❑ **For Orfadin suspension requests:** Provider must submit documentation to confirm the member is unable to swallow nitisinone capsules **(submit documentation of intolerance to capsules)**
- ❑ **For Harliku requests:** Member must have trial and failure of **BOTH** of the following unless contraindicated or clinically significant adverse effects are experienced **[verified by chart notes and/or pharmacy paid claims. Provider must submit documentation as to why the member cannot be prescribed a preferred agent. Include details and a completed FDA MedWatch Form (https://www.accessdata.fda.gov/scripts/medwatch/index.cfm) is required to be attached for adverse reactions to prerequisite therapies.]**
  - ❑ Generic nitisinone capsules or Brand Orfadin capsules/suspension ***requires prior authorization***
  - ❑ Nityr ***requires prior authorization***
- ❑ Member is **NOT** using two different nitisinone products concurrently
- ❑ Prescribed dose does not exceed 2 mg per day

Reauthorization: 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.

- ❑ Member continues to meet all initial authorization criteria
- ❑ Member is responding positively to therapy as evidenced by at least **ONE** of the following **(check all that apply, submit documentation)**:
  - ❑ Reduced levels of urinary HGA
  - ❑ Improved joint (e.g., hip, spine, knee, shoulder) symptoms (e.g., range of motion, pain, stiffness)

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Medication being provided by Specialty Pharmacy – Proprium Rx

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.****

***Approved by Pharmacy and Therapeutics Committee**: ~~11/21/2019;~~ 9/25/2025
REVISED/UPDATED/REFORMATTED: ~~2/25/2020; 10/11/2021; 11/1/2023;~~ **10/22/2025**