JOHNS HOPKINS MEDICINE JOHNS HOPKINS HEALTHCARE	Johns Hopkins HealthCare LLC **Pharmacy Public** **Pharmacy Management Drug Policies**	*Policy Number*	MEDS129
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	Subject **Orfadin, Nityr, nitisinone**	*Revision Date*	12/08/2021
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This document applies to the following Participating Organizations:

Priority Partners

Keywords: Nitisinone , Nityr, Orfadin

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I. POLICY

A. Orfadin (nitisinone) capsules and oral suspension, and Nityr (nitisinone) tablets will require prior authorization to ensure appropriate use. The process for initiating a prior authorization request can be found in policy PHARM 20.
   1. PPMCO members are subject to the Priority Partners formulary, available at www.ppmco.org.
   2. USFHP members are subject to prior authorization criteria, step-edits and days-supply limits outlined in the Tricare Policy Manual. Tricare Policy supersedes JHHC Medical/Pharmacy Policies. Tricare limits may be accessed at: http://pec.ha.osd.mil/formulary_search.php?submenuheader=1

II. POLICY CRITERIA

A. **Nitisinone capsules (generic of Orfadin capsules)** and **Orfadin oral suspension** may be approved for patients meeting the following:
   1. Documented diagnosis of hereditary tyrosinemia type 1(HT-1)
   2. Confirmation of diagnosis with laboratory testing showing detection of succinylacetone in urine or plasma, or generic testing
   3. Documentation showing that the requested medication will be used as an adjunct to dietary restriction of tyrosine and phenylalanine

B. Brand **Orfadin capsules**, and **Nityr tablets** may be approved for patients meeting the following:
   1. Documented diagnosis of hereditary tyrosinemia type 1(HT-1)
   2. Confirmation of diagnosis with laboratory testing showing detection of succinylacetone in urine or plasma, or generic testing
   3. Documentation showing that the requested medication will be used as an adjunct to dietary restriction of tyrosine and phenylalanine
   4. Documented trial and inadequate response or intolerance with generic nitisinone capsules

III. AUTHORIZATION PERIOD/LIMITATIONS

A. Initial approval will be for 12 months of therapy

B. Approval for continuation of therapy may be extended in 12-month intervals with documentation showing beneficial response to treatment.

IV. EXCLUSIONS

A. Nitisinone products will not be approved for the following:
   1. Doses exceeding the maximum total daily dosage of 2mg/kg
   2. Any indications or usage that is not FDA-approved, or guideline-supported

B. The use of physician samples, or manufacturer product discounts, does not guarantee coverage under the provisions of the medical and/or pharmacy benefit. All pertinent criteria must be met in order to be eligible for benefit coverage.

V. REFERENCES

1. Orfadin [prescribing information]. Waltham, MA: Sobi Inc; May 2019
2. Nityr [prescribing information]. Cambridge, United Kingdom: Cycle Pharmaceuticals Ltd.; October 2020

VI. APPROVALS

Signature on file at JHHC

DATE OF REVISION	SUMMARY OF CHANGE
01/20/2021	Policy Creation
12/08/2021	Updated Exclusions section regarding physician samples

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