STANDARD COMMERCIAL DRUG FORMULARY
PRIOR AUTHORIZATION GUIDELINES

NITISINONE

Generic	Brand	HICL	GCN	Medi-Span	Exception/Other
NITISINONE	ORFADIN, NITYR, NITISINONE	23253		GPI-10 (3090404500)	

GUIDELINES FOR USE

INITIAL CRITERIA (NOTE: FOR RENEWAL CRITERIA SEE BELOW)

1. Does the patient have a documented diagnosis of hereditary tyrosinemia type 1 (HT-1) and meet **ALL** of the following criteria?
   - The patient has elevated urinary or plasma succinylacetone (SA) levels OR a mutation in the fumarylacetoacetate hydrolase (FAH) gene
   - Therapy is prescribed by or in consultation with a prescriber specializing in inherited metabolic diseases
   - The patient has been counseled on maintaining dietary restriction of tyrosine and phenylalanine

   If yes, continue to #2.
   If no, do not approve.
   DENIAL TEXT: See the initial denial text at the end of the guideline.

2. Is the request for Nityr tablets; brand Orfadin 2mg, 5mg, 10 mg, 20 mg capsules; or Orfadin suspension **AND** the patient meets the following criterion?
   - The patient had a trial of or contraindication to generic nitisinone capsule

   If yes, **approve the requested drug for 6 months by GPID or GPI-14.**
   If no, do not approve.
   DENIAL TEXT: See the initial denial text at the end of the guideline.

CONTINUED ON NEXT PAGE

NITISINONE

INITIAL CRITERIA (CONTINUED)

INITIAL DENIAL TEXT: *Some terms are already pre-defined in parenthesis. Please use these definitions if the particular text you need to use does not already have definition(s) in it.

Our guideline named **NITISINONE (Orfadin, Nityr)** requires the following rule(s) be met for approval:

A. You have hereditary tyrosinemia type 1 (HT-1: a type of genetic disorder where you cannot breakdown an important component in proteins)
B. Your diagnosis is confirmed by elevated urinary or plasma succinylacetone levels (a chemical that is present in hereditary tyrosinemia) OR a mutation in the fumarylacetoacetate hydrolase gene
C. Therapy is prescribed by or in consultation with a prescriber specializing in inherited metabolic diseases
D. You have been counseled on maintaining dietary restriction of tyrosine and phenylalanine
E. **If you are requesting Nityr tablets; brand Orfadin 2mg, 5mg, 10 mg, 20 mg capsules; or Orfadin oral suspension, approval also requires:**
    1. You have tried or have a contraindication (harmful for) to generic nitisinone capsules

Your doctor told us [INSERT PT SPECIFIC INFO PROVIDED]. We do not have information showing you [INSERT UNMET CRITERIA]. This is why your request is denied. Please work with your doctor to use a different medication or get us more information if it will allow us to approve this request.

RENEWAL CRITERIA

1. Does the patient have a diagnosis of hereditary tyrosinemia type 1 **AND** meet the following criterion?
    - The patients urinary or plasma succinylacetone (SA) levels have decreased from baseline while on treatment with nitisinone.

    If yes, **approve for 12 months by GPID or GPI-14 for all strengths of the requested formulation.**
    If no, do not approve.
    DENIAL TEXT: See the renewal denial text at the end of the guideline.

CONTINUED ON NEXT PAGE

NITISINONE

RENEWAL CRITERIA (CONTINUED)

RENEWAL DENIAL TEXT: *Some terms are already pre-defined in parenthesis. Please use these definitions if the particular text you need to use does not already have definition(s) in it.

Our guideline named **NITISINONE (Orfadin, Nityr)** requires the following rule(s) be met for renewal:

A. You have hereditary tyrosinemia type 1 (HT-1: a type of genetic disorder where you cannot breakdown an important component in proteins)
B. Your urinary or plasma succinylacetone levels (a chemical that is present in hereditary tyrosinemia) have decreased from baseline while on treatment with nitisinone

Your doctor told us [INSERT PT SPECIFIC INFO PROVIDED]. We do not have information showing you [INSERT UNMET CRITERIA]. This is why your request is denied. Please work with your doctor to use a different medication or get us more information if it will allow us to approve this request.

RATIONALE

For further information, please refer to the Prescribing Information and/or Drug Monograph for Orfadin and Nityr.

REFERENCES

- Orfadin [Prescribing Information]. Waltham, MA: Sobi, Inc.; May 2019.
- Nityr [Prescribing Information]. Cambridge, UK: Cycle Pharmaceuticals Ltd.; September 2020.

Library	Commercial	NSA
Yes	Yes	No

Part D Effective: N/A
Commercial Effective: 04/17/23
Created: 08/16
Client Approval: 03/23
P&T Approval: 07/18