Pharmacy Medical Necessity Guidelines: Nityr™ and Orfadin® (nitisinone)

Effective: December 17, 2018

Prior Authorization Required √ Type of Review – Care Management
Not Covered Type of Review – Clinical Review √
Pharmacy (RX) or Medical (MED) Benefit Rx Department to Review RxUM

These pharmacy medical necessity guidelines apply to the following:

Commercial Products
☑ Tufts Health Plan Commercial products – large group plans
☑ Tufts Health Plan Commercial products – small group and individual plans
☑ Tufts Health Freedom Plan products – large group plans
☑ Tufts Health Freedom Plan products – small group plans
• CareLinkSM – Refer to CareLink Procedures, Services and Items Requiring Prior Authorization

Tufts Health Public Plans Products
☑ Tufts Health Direct – A Massachusetts Qualified Health Plan (QHP) (a commercial product)
☑ Tufts Health Together – MassHealth MCO Plan and Accountable Care Partnership Plans
☑ Tufts Health RITogether – A Rhode Island Medicaid Plan

Note: This guideline does not apply to Medicare Members (includes dual eligible Members).

OVERVIEW
Hereditary tyrosinemia type 1 (HT-1) is a rare genetic metabolic disorder that causes progressive liver disease and renal tubular dysfunction. The disorder is caused by fumarylacetoacetate hydrolase deficiency, the last enzyme in the degradation pathway of the amino acid tyrosine, resulting in the accumulation of toxic metabolites. HT-1 is present at birth and manifests within weeks or months in the failure of the infant to thrive and by symptoms of hepatomegaly, edema, ascites, melena, and hemorrhagic diathesis.

Nityr and Orfadin (nitisinone) inhibit 4-hydroxyphenylpyruvate dioxygenase and prevents the formation of toxic metabolites involved in hepatic and renal lesions. Because these medications inhibit the breakdown of tyrosine which leads to increase plasma levels of tyrosine, patients receiving therapy must restrict their dietary intake of tyrosine and phenylalanine.

FOOD AND DRUG ADMINISTRATION-APPROVED INDICATIONS
Nityr and Orfadin (nitisinone) are indicated for the treatment of HT-1 in combination with dietary restriction of tyrosine and phenylalanine.

COVERAGE GUIDELINES
The plan may authorize coverage of Orfadin or Nityr (nitisinone) when the following criterion is met:
1. Documentation of genetic tyrosinemia Type-1 (hereditary tyrosinemia Type-1)

LIMITATIONS
• Orfadin and Nityr (nitisinone) will not be covered for any indication other than tyrosinemia Type-1 (hereditary tyrosinemia Type-1).

CODES
None

REFERENCES

APPROVAL HISTORY
November 2002: Reviewed by Pharmacy & Therapeutics Committee.
Subsequent endorsement date(s) and changes made:
1. August 9, 2005: No changes
2. September 14, 2004: No changes
3. July 11, 2006: No changes
4. July 10, 2007: No changes
5. July 08, 2008: No changes
6. July 14, 2009: No changes
7. January 1, 2010: Removal of Tufts Medicare Preferred language (separate criteria have been created specifically for Tufts Medicare Preferred)
8. July 13, 2010: No changes
9. July 12, 2011: No changes
10. April 10, 2012: No changes
11. March 12, 2013: No changes
12. March 11, 2014: No changes
13. March 10, 2015: No changes
15. February 9, 2016: No changes
16. July 12, 2016: No changes
18. July 11, 2017: No changes
19. December 12, 2017: Changed the name of the Medical Necessity Guideline to Orfadin and Nityr (nitisinone). Added Nityr (nitisinone) to the Medical Necessity Guideline. Effective 12/18/17, the Medical Necessity Guideline applies to Tufts Health Together and Tufts Health RITogether.

BACKGROUND, PRODUCT AND DISCLAIMER INFORMATION

Pharmacy Medical Necessity Guidelines have been developed for determining coverage for plan benefits and are published to provide a better understanding of the basis upon which coverage decisions are made. The plan makes coverage decisions on a case-by-case basis considering the individual member's health care needs. Pharmacy Medical Necessity Guidelines are developed for selected therapeutic classes or drugs found to be safe, but proven to be effective in a limited, defined population of patients or clinical circumstances. They include concise clinical coverage criteria based on current literature review, consultation with practicing physicians in the service area who are medical experts in the particular field, FDA and other government agency policies, and standards adopted by national accreditation organizations. The plan revises and updates Pharmacy Medical Necessity Guidelines annually, or more frequently if new evidence becomes available that suggests needed revisions.

For self-insured plans, coverage may vary depending on the terms of the benefit document. If a discrepancy exists between a Pharmacy Medical Necessity Guideline and a self-insured Member’s benefit document, the provisions of the benefit document will govern.

Treating providers are solely responsible for the medical advice and treatment of members. The use of this policy is not a guarantee of payment or a final prediction of how specific claim(s) will be adjudicated. Claims payment is subject to member eligibility and benefits on the date of service, coordination of benefits, referral/authorization and utilization management guidelines when applicable, and adherence to plan policies and procedures and claims editing logic.