# Criteria-Based Consultation Prescribing Program
# CRITERIA FOR DRUG COVERAGE
# Nitisinone capsule (Orfadin)

**Notes**:

- * Intolerance excludes adverse drug reactions that are expected, mild in nature, resolve with continued treatment, and do not require medication discontinuation

**Initiation (new start) criteria:** Non-formulary **nitisinone capsule (Orfadin)** will be covered on the prescription drug benefit for 12 months when the following criteria are met:

- Prescriber is a geneticist or a metabolic specialist.
- Patient has a diagnosis of hereditary tyrosinemia type-1 (HT-1) confirmed by one of following:
    i. Elevated succinylacetone levels in blood or urine samples
    ii. DNA testing: mutation in the fumarylacetoacetate hydrolase (FAH) gene

  **-OR-**

  Patient has a diagnosis of alkaptonuria (AKU), evidenced by high levels of homogentisic acid (HGA) in the urine.
- Nitisinone is used in conjunction with a tyrosine and phenylalanine diet restriction when used for HT-1 treatment

**Criteria for *current Kaiser Permanente members* already taking the medication who have not been reviewed previously:** Non-formulary **nitisinone capsule (Orfadin)** will be covered on the prescription drug benefit for 12 months when the following criteria are met:

- Prescriber is a geneticist or a metabolic specialist.
- Patient has a diagnosis of hereditary tyrosinemia type-1 (HT-1) confirmed by one of following:
    i. Elevated succinylacetone levels in blood or urine samples
    ii. DNA testing: mutation in the fumarylacetoacetate hydrolase (FAH) gene

  **-OR-**

  Patient has a diagnosis of alkaptonuria (AKU), evidenced by high levels of homogentisic acid (HGA) in the urine.
- Nitisinone is used in conjunction with a tyrosine and phenylalanine diet restriction when used for HT-1 treatment

# Criteria-Based Consultation Prescribing Program
# CRITERIA FOR DRUG COVERAGE
# Nitisinone capsule (Orfadin)

**Criteria for *new members* entering Kaiser Permanente already taking the medication who have not been reviewed previously:** Non-formulary **nitisinone capsule (Orfadin)** will be covered on the prescription drug benefit for 12 months when the following criteria are met:

- Prescriber is a geneticist or a metabolic specialist.
- Patient has a diagnosis of hereditary tyrosinemia type-1 (HT-1) confirmed by one of following:
    i. Elevated succinylacetone levels in blood or urine samples
    ii. DNA testing: mutation in the fumarylacetoacetate hydrolase (FAH) gene

    **-OR-**

    Patient has a diagnosis of alkaptonuria (AKU), evidenced by high levels of homogentisic acid (HGA) in the urine.
- Nitisinone is used in conjunction with a tyrosine and phenylalanine diet restriction when used for HT-1 treatment

**Continued use criteria 12 months after initiation):** Non-formulary **nitisinone (Orfadin)** will continue to be covered on the prescription drug benefit when the following criteria are met:

- Prescriber continues to be a geneticist or a metabolic specialist.
- Patient continues tyrosine and phenylalanine dietary restriction when used for HT-1 treatment
- Patient is receiving clinical benefit to nitisinone as indicated by decreased succinylacetone, or alpha-1-microglobulin, or urine homogentisic acid (HGA) levels