

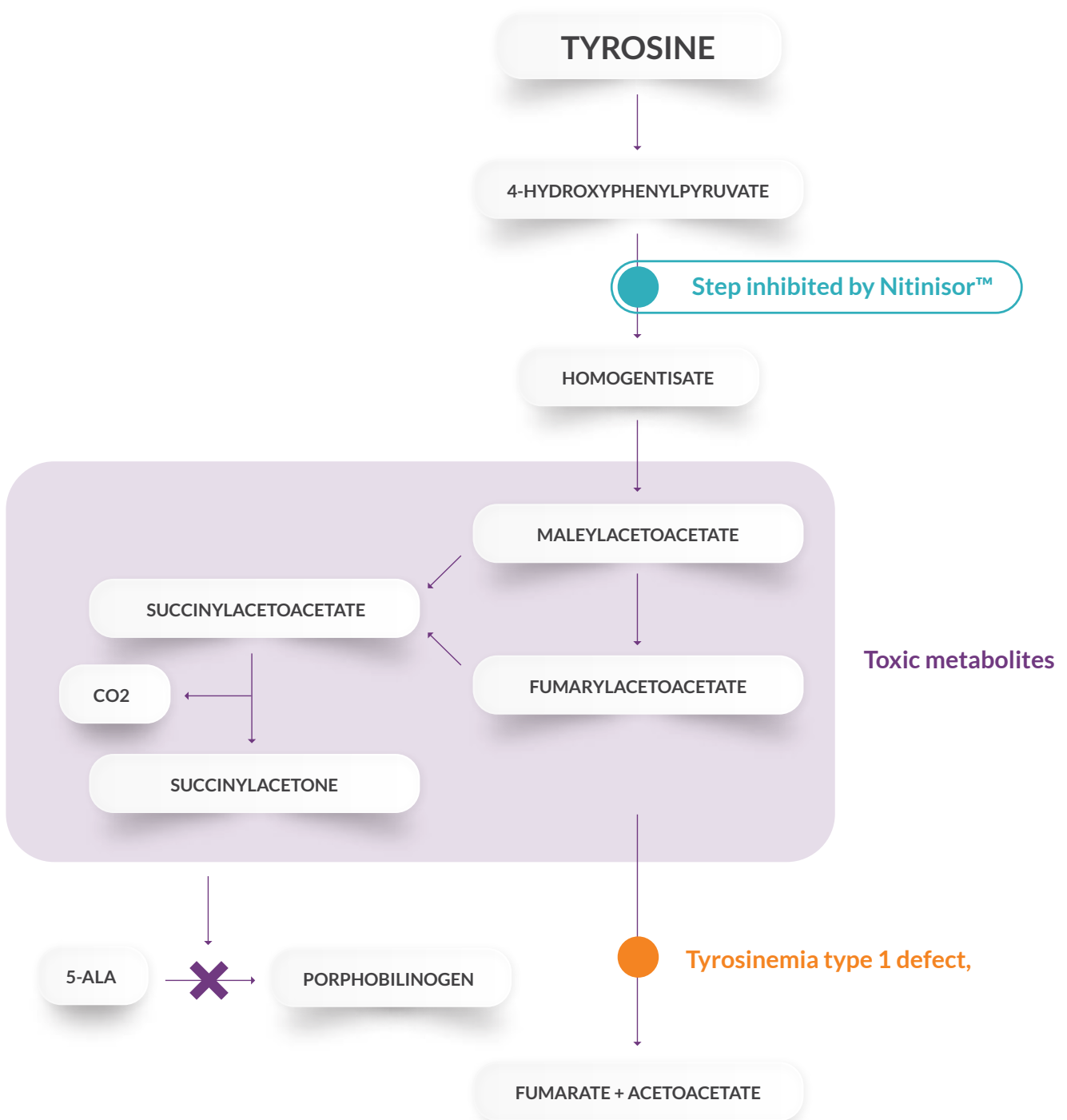
NitinisorTM

(Nitisinone)

Summary of Product Characteristics



Tyrosinemia type 1 is a rare, congenital metabolic disorder of tyrosine metabolism, resulting in the accumulation of toxic metabolites that damage the liver and kidneys. The illness classically presents as severe liver disease in young infants. Children older than six months may come to medical attention with signs of rickets or neurologic crises.



Nitisor™

(Nitisonone)

By inhibiting the normal catabolism of tyrosine in patients with tyrosinemia type 1, Nitisor™ prevents the accumulation of the toxic metabolites that cause liver and kidney damage.

Since 1991, when the first patient started treatment with nitisonone.¹

Treatment of all genotypes of the disease should be initiated as early as possible to increase overall survival and avoid complications such as liver failure, liver cancer and renal disease.²

The recommended dose is 1-2 mg/kg bodyweight per day. When the dose is properly adjusted, succinylacetone should not be detectable in urine or plasma.²

References

1. Internal communication.
2. Summary of Product Characteristics, Nitisor™.