Xenazine®
Covered Medication

- Tetrabenazine (Xenazine®)

What it does and how it is used
- Huntington’s disease is a rare inherited disease that affects approximately 15 of 100,000 individuals in the US. It is progressive, degenerative, eventually fatal, and is characterized by hyperkinetic movement disorders and degeneration in mental and emotional capacities.
- Chorea, an involuntary movement disorder consisting of non-repetitive, rapid, ceaseless jerking of limbs, face, or trunk, is the major sign of the disease, and presents in up to 90% of all Huntington’s patients.
- This movement disorder results from a degeneration of the cells of the basal ganglia of the brain and differs from the repetitive tremors seen in patients with Parkinson’s Disease whose symptoms occur due to a deficiency of dopamine.
- Currently, there are no measures available to control the disease progression itself. All treatment options are directed at controlling the disease symptoms, such as antidepressants, antipsychotics, tranquilizers, and mood stabilizers to treat depression, psychosis, cognitive impairments, irritability, rigidity, insomnia, obsessive compulsive disorder, apathy, and chorea.
- Tetrabenazine® is used for the symptomatic treatment of hyperkinetic movement disorders associated with Huntington’s chorea. Tetrabenazine is also used in other countries for treatment of various other movement disorders not associated with Huntington’s disease, such as Tourette’s syndrome and tardive dyskinesia.
- Although it is unknown exactly how Tetrabenazine® works to reduce chorea symptoms, it is believed to be related to its effect as a reversible depletor of monoamines, such as dopamine, from nerve terminals. Unlike reserpine, another monoamine depletor, Tetrabenazine® is specific for functions in areas of the CNS where the pathology of the disease originates.
- To quantify the degree of chorea experienced by Huntington’s patients, the Huntington Study Group utilized the Unified Huntington’s Disease Rating Scale (UHDRS), which is a system based on maximum achieved scores to assess the severity of the condition. The efficacy of Tetrabenazine® is definitive in that it’s been shown to reduce UHDRS maximal chorea score in comparison with placebo, and that there is a significant rate of reemergence of chorea upon discontinuation of Tetrabenazine®.
- The daily dose of Tetrabenazine® should be individualized.
- The recommended dose for Tetrabenazine® is 12.5 mg per day initially given once in the morning, and then increased after one week to 12.5 mg twice a day; subsequent dose titration should increase the daily dose by 12.5 mg every week until an optimal dose is reached that is well tolerated and controls chorea.
- CYP 2D6 is the major metabolism pathway that contributes to the elimination of Tetrabenazine®. The excretion of Tetrabenazine® is slower in patients with poor CYP 2D6 function, when compared to patients with normal to extensive CYP 2D6 function.
- Genotyping of CYP 2D6 should be performed for all patients expected to need therapeutic dose above 50 mg per day. Poor metabolizers of 2D6 should not receive daily doses exceeding 50 mg per day due to decreased drug clearance and increased drug toxicity; extensive and intermediate metabolizers of 2D6 may receive doses up to 100 mg per day.
- Treatment with Tetrabenazine® may be discontinued without tapering.
- If the patient does not experience improvement after seven days of therapy on maximum tolerated dose, it is unlikely that Tetrabenazine® will be of benefit to the patient, either by increasing dose or by extending the duration of treatment.

What it costs
- AWP is $37.50 per 12.5 mg & $74.50 per 25 mg tablet of Tetrabenazine®, both available in bottles of 112 tablets.
- $4,470 per month at 50 mg daily, or $53,640 per year

Rationale for coverage authorization
To reduce exposure to costs associated with uses of Tetrabenazine® other than for chorea associated with Huntington’s disease, and to provide coverage for dosages up to 100 mg per day.

Benefit design
Coverage for Tetrabenazine® is determined through a prior authorization process for every claim AND Coverage is provided for a quantity sufficient for dosing up to 100mg per day.
## Coverage authorization criteria

Coverage for Xenazine® is provided through prior authorization in accord with the following:

1. Coverage is provided for the treatment of chorea associated with Huntington’s disease.

**Coverage duration:** Coverage is provided for the lifetime of the request for a quantity not to exceed 100 mg per day.

## References