DISCLAIMER: This template of appeal and/or medical necessity, which should be sent on the Healthcare Provider's (HCP) letterhead, is available to all HCPs and clinicians treating patients diagnosed with Wilson Disease for whom they want to prescribe ***CUVRIOR® (trientine tetrahydrochloride).*** *Orphalan employees cannot assist with the completion of this template letter or customize it in any way. HCPs/Clinicians, however, may use this template to provide patient specific clinical information to the applicable payer and must determine the accuracy of all statements therein. This template letter may be used regardless of payer plan or type, but Orphalan makes no representation or guarantee of the outcome or expected results of the use of this template.*

[Name of Insurance Company] [Date]
[Address of Insurance Company]
[City, State, ZIP of Insurer]

Re: Statement of [Appeal and/or Medical Necessity] for CUVRIOR® (trientine tetrahydrochloride) for treatment of Wilson Disease.

[Patient Name]
[Name of Policyholder]
[Patient Date of Birth}
[Insurance ID Number of Patient]
[Insurance Group Number of Patient]

To Whom It May Concern:

I am writing on behalf of my patient, [Patient's name], to document the [medical necessity and/or appeal] for the use of CUVRIOR for Wilson Disease [ICD-10 code].

[Patient's name] was diagnosed with Wilson Disease on [original diagnosis date]. [Patient's name] has experienced [add any recent symptoms, conditions, treatments affecting the patient's struggle with changes in quality-of-life assessment, activities of daily living affected by current Wilson Disease treatment. (*Omit If nothing pertinent*.)]

This patient has previously received the following treatment(s) for Wilson Disease: [indicate which prior therapies the patient has received, including all Wilson Disease related medications, and outline the specific reasons for any discontinuation or non- compliance].

Treatment with CUVRIOR is medically necessary because of [Choose any or all of the following in your judgment (or add additional reasons based on your clinical judgment): adverse effects and/or poor quality of life (with or without progression)]. Additionally, [insert statement(s), if applicable to patient: Cuvrior provides convenient twice-daily dosing with no refrigeration for long term maintenance therapy. Cuvrior tablets are scored and can be divided in half, allowing for flexible and accurate dosing]. Thus, I believe that treatment with CUVRIOR is medically necessary and will provide essential clinical benefit in [patient name]'s current course of care.

Please do not hesitate to contact me at [physician's telephone number] if you require any further information to approve this request.

Thank you for an expedient response,

[Physician Signature]

[Physician Name, M.D.]
[Practice Name]

CUVRIOR® (trientine tetrahydrochloride) Tablets

INDICATION

CUVRIOR is a copper chelator indicated for the treatment of adult patients with stable Wilson's disease who are de-coppered and tolerant to penicillamine.

IMPORTANT SAFETY INFORMATION

CONTRAINDICATIONS

- CUVRIOR is contraindicated in patients with hypersensitivity to trientine or to any of the excipients in CUVRIOR.

WARNINGS AND PRECAUTIONS

- **Potential for Worsening of Clinical Symptoms at Initiation of Therapy**, including neurological deterioration, may occur at the beginning of CUVRIOR therapy due to mobilization of excess stores of copper. Adjust the dosage or discontinue therapy if clinical condition worsens. Evaluate serum non-ceruloplasmin copper (NCC) levels or 24-hour urinary copper excretion (UCE) when initiating treatment, after 3 months, and approximately every 6 months thereafter.
- **Copper Deficiency** may develop following treatment with CUVRIOR. Periodic monitoring is required.
- **Iron Deficiency** may develop following treatment with CUVRIOR. If iron deficiency develops, a short course of iron supplementation may be given.
- **Hypersensitivity Reactions**, characterized by rash, have been reported with the use of trientine. Rash was reported in 12% (3/26) of CUVRIOR-treated patients, and one patient discontinued treatment due to rash. If rash or other hypersensitivity reaction occurs, consider discontinuing CUVRIOR.

ADVERSE REACTIONS

The most common adverse reactions (>5%) are abdominal pain, change of bowel habits, rash, alopecia, and mood swings.

DRUG INTERACTIONS

- **Mineral Supplements** (e.g. iron, zinc, calcium, magnesium): Avoid concomitant use. If concomitant use is unavoidable, take CUVRIOR at least 2 hours before or 2 hours after iron and take CUVRIOR at least 1 hour before or 2 hours after other mineral supplements.
- **Other Drugs for Oral Administration:** Take CUVRIOR at least 1 hour apart from any other oral drug.

Please see full Prescribing Information at www.cuvrior.com.

To report SUSPECTED ADVERSE REACTIONS, contact Orphalan at 1-800-961-8320 or FDA at 1-800-FDA-1088 or www.fda.gov/medwatch.