Policy and Procedure

DEPARTMENT: Pharmacy, Medical Directors	**DOCUMENT NAME:** cerliponase alfa (Brineura®)
PAGE: 1 of 4	**REPLACES DOCUMENT:**
APPROVED DATE: 4/6/2018	**RETIRED:**
EFFECTIVE DATE: 4/6/2018	**REVIEWED/REVISED:** 2/13/2019, 1/6/20
PRODUCT TYPE: Star, Star Health, Star Kids, Star Plus, Chip, Chip Prenate	**REFERENCE NUMBER:** TX.PHAR.46

SCOPE:

Superior Health Plan Pharmacy Department, Medical Directors

PURPOSE:

It is the policy of Superior HealthPlan to follow state guidance for medical necessity review of cerliponase alfa (Brineura®). This medication is a pass through drug and should follow state guidance for medical necessity review for Medicaid/CHIP due to the manner in which it is reimbursed. All determinations will be performed by a Superior Medical Director. A pharmacy clinician will review the prior authorization request and make a recommendation to the Medical Director but will not make the ultimate determination on any case.

BACKGROUND:

Description:
Cerliponase alfa (Brineura®) is a hydrolytic lysosomal N-terminal tripeptidyl peptidase.

FDA Approved Indication(s)
Brineura® is indicated to slow the loss of ambulation in symptomatic pediatric patients 3 years of age and older with late infantile neuronal ceroid lipofuscinosis type 2 (CLN2), also known as tripeptidyl peptidase 1 (TPP1) deficiency.

Formulations:
Injection: Brineura® 150 mg/5 mL (30 mg/mL) solution, two single-dose vials per carton co-packaged with Intraventricular Electrolytes Injection 5 mL in a single-dose vial.

Policy and Procedure

DEPARTMENT: Pharmacy, Medical Directors	**DOCUMENT NAME:** cerliponase alfa (Brineura®)
PAGE: 2 of 4	**REPLACES DOCUMENT:**
APPROVED DATE: 4/6/2018	**RETIRED:**
EFFECTIVE DATE: 4/6/2018	**REVIEWED/REVISED:** 2/13/2019, 1/6/20
PRODUCT TYPE: Star, Star Health, Star Kids, Star Plus, Chip, Chip Prenate	**REFERENCE NUMBER:** TX.PHAR.46

PROCEDURE:

Provider must submit documentation (which may include office chart notes and lab results) supporting that member has met all approval criteria.

I. Initial Approval Criteria:

1. A Medical Director is required to review and approve or deny all requests.
2. Diagnosis of late infantile neuronal CLN2;
3. Confirmation of CLN2 with both of the following:
   a. TPP1 enzyme activity test demonstrating deficient TPP1 enzyme activity in leukocytes;
   b. Identification of 2 pathogenic mutations *in trans* in the TPP1/CLN2 gene.
4. Prescribed by or in consultation with a neurologist;
5. Motor domain of the CLN2 Clinical Rating Scale score ≥ 1;
6. Dose does not exceed 300 mg administered once every other week as an intraventricular infusion;
7. At the time of request, member does not have ventriculoperitoneal shunts.

Approval duration: 6 months

II. Continued Therapy:

1. A Medical Director is required to review and approve or deny all continuation requests.
2. Currently receiving medication via Centene benefit, or member has previously met initial approval criteria or was on the therapy by another managed care organization.
3. Diagnosis of late infantile neuronal CLN2.
4. Documentation of positive response to therapy defined as no decline or decline of only one category and a score > 0 on the CLN2 Clinical Rating Scale;
5. If request is for a dose increase, new dose does not exceed 300 mg administered once every other week as an intraventricular infusion.

DEPARTMENT: Pharmacy, Medical Directors	**DOCUMENT NAME:** cerliponase alfa (Brineura®)
PAGE: 3 of 4	**REPLACES DOCUMENT:**
APPROVED DATE: 4/6/2018	**RETIRED:**
EFFECTIVE DATE: 4/6/2018	**REVIEWED/REVISED:** 2/13/2019, 1/6/20
PRODUCT TYPE: Star, Star Health, Star Kids, Star Plus, Chip, Chip Prenate	**REFERENCE NUMBER:** TX.PHAR.46

Approval duration: 6 months

III. Appendices/General Information

Appendix: Motor Domain of CLN2 Clinical Rating Scale

- The motor domain of the CLN2 Clinical Rating Scale is scored as follow: walks normally = 3, intermittent falls, clumsiness, obvious instability = 2, no unaided walking or crawling only = 1, immobile, mostly bedridden = 0.
- Decline was defined as having an unreversed (sustained) 2 category decline or an unreversed score of 0 in the motor domain of the CLN2 Clinical Rating Scale.

IV. Dosage and Administration

Indication	Dosing Regimen	Maximum Dose
CLN2	300 mg administered once every other week as an intraventricular infusion followed by infusion of intraventricular electrolytes over approximately 4.5 hours	300 mg every other week

REFERENCES:

ATTACHMENTS:

DEFINITIONS/Abbreviations:

CLN2: ceroid lipofuscinosis type 2

Policy and Procedure

DEPARTMENT: Pharmacy, Medical Directors	**DOCUMENT NAME:** cerliponase alfa (Brineura®)
PAGE: 4 of 4	**REPLACES DOCUMENT:**
APPROVED DATE: 4/6/2018	**RETIRED:**
EFFECTIVE DATE: 4/6/2018	**REVIEWED/REVISED:** 2/13/2019, 1/6/20
PRODUCT TYPE: Star, Star Health, Star Kids, Star Plus, Chip, Chip Prenate	**REFERENCE NUMBER:** TX.PHAR.46

FDA: Food and Drug Administration
TPP1: tripeptidyl peptidase 1

REVISION LOG

REVISION	DATE
Changed "Justin M. Weiss, Sr. V.P., Pharmacy Operations" to "Karen Tadlock, V.P., Pharmacy Operations" Formatting	2/13/2019
Revised wording/clarified requirement for step #4 under Continued Therapy criteria Formatting	1/6/2020

POLICY AND PROCEDURE APPROVAL

Karen Tadlock, V.P., Pharmacy Operations — Approval on file

Dr. David Harmon, Sr. V.P., Chief Medical Officer — Approval on file

Pharmacy & Therapeutics Committee: — Approval on file

NOTE: The electronic approval retained in RSA Archer, Centene's P&P management software, is considered equivalent to a physical signature.