Policy Title:	Brineura (cerliponase alfa) (Intravenous)		
		Department:	PHA
Effective Date:	01/01/2020		
Review Date:	12/4/2019		
Revision Date:	12/4/2019		

Purpose: To support safe, effective and appropriate use of Brineura (cerliponase alfa).

Scope: Medicaid, Exchange, Medicare-Medicaid Plan (MMP)

Policy Statement:

Brineura (cerliponase alfa) is covered under the Medical Benefit when used within the following guidelines. Use outside of these guidelines may result in non-payment unless approved under an exception process.

Procedure:

Coverage of Brineura (cerliponase alfa) will be reviewed prospectively via the prior authorization process based on criteria below.

Initial Criteria:

- Patient has one of the following diagnosis:
  - Diagnosis of late infantile neuronal ceroid lipofuscinosis type 2 (CLN2) by a neurologist with expertise in the diagnosis of CLN2
  - Diagnosis of late infantile neuronal ceroid lipofuscinosis type 2 (CLN2) by a physician in consultation with a neurologist with expertise in the diagnosis of CLN2; AND
- Patient is age 3 years or older; AND
- All of the following scores on the Clinical Scoring System for Late Infantile Neuronal Ceroid Lipofuscinosis (LINCL):
  - Combined score of 3 to 6 in the motor and language domains
  - Score of at least 1 in the motor domain
  - Score of at least 1 in the language domain; AND
- Brineura is prescribed by a neurologist with expertise in the treatment of CLN2 or Brineura is prescribed by a physician in consultation with a neurologist with expertise in the treatment of CLN2; AND
- Brineura is to be administered intraventricularly by, or under the direction of, healthcare professionals experienced in performing intraventricular infusions via an intracerebroventricular catheter; AND

- Dosing is in accordance with the United States Food and Drug Administration approved labeling: 300 mg administered once every other week as an intraventricular infusion

Continuation of Therapy Criteria:

- Patient is tolerating treatment; AND
- Patient has one of the following diagnosis:
    - Diagnosis of late infantile neuronal ceroid lipofuscinosis type 2 (CLN2) by a neurologist with expertise in the diagnosis of CLN2
    - Diagnosis of late infantile neuronal ceroid lipofuscinosis type 2 (CLN2) by a physician in consultation with a neurologist with expertise in the diagnosis of CLN2; AND
- Patient is age 3 years or older; AND
- Patient has a score of 1 or higher in the motor domain of the Clinical Scoring System for LINCL; AND
- Brineura is prescribed by a neurologist with expertise in the treatment of CLN2 or Brineura is prescribed by a physician in consultation with a neurologist with expertise in the treatment of CLN2; AND
- Brineura is to be administered intraventricularly by, or under the direction of, healthcare professionals experienced in performing intraventricular infusions via an intracerebroventricular catheter; AND
- Dosing is in accordance with the United States Food and Drug Administration approved labeling: 300 mg administered once every other week as an intraventricular infusion

Coverage durations:

- Initial coverage: 6 months
- Continuation of therapy coverage: 6 months

*** Requests will also be reviewed to National Coverage Determination (NCD) and Local Coverage Determinations (LCDs) if applicable.***

Dosage/Administration:

Indication	Dose	Maximum dose (1 billable unit = 1 mg)
CLN2	300 mg administered once every other week by intraventricular infusion	300 billable units (2 kits) every 14 days

Investigational use: All therapies are considered investigational when used at a dose or for a condition other than those that are recognized as medically accepted indications as defined in any one of the following standard reference compendia: American Hospital Formulary Service Drug information (AHFS-DI), Thomson Micromedex DrugDex, Clinical Pharmacology, Wolters Kluwer Lexi-Drugs, or Peer-reviewed published medical literature indicating that sufficient evidence exists to support use. Neighborhood does not provide coverage for drugs when used for investigational purposes.

Applicable Codes:
Below is a list of billing codes applicable for covered treatment options. The below tables are provided for reference purposes and may not be all inclusive. Requests received with codes from tables below do not guarantee coverage. Requests must meet all criteria provided in the procedure section.

The following HCPCS/CPT codes are:

HCPCS/CPT Code	Description
J0567	Injection, cerliponase alfa, 1 mg:

References:

1. Brineura [prescribing information]. Novato, CA: BioMarin Pharmaceutical Inc.; December 2018.
2. Williams RE, Adams HR, Blohm M, Cohen-Pfeffer JL, de Los Reyes E, Denecke J, et al. Management Strategies for CLN2 Disease. Pediatr Neurol. 2017 Apr;69:102-112.
3. http://www.cln2connection.com/overview/cln2-disease. Accessed May 30, 2019.
4. Steinfeld R, Heim P, von Gregory H, et al. Late infantile neuronal ceroid lipofuscinosis: quantitative description of the clinical course in patients with CLN2 mutations. Am J Med Genet. 2002;112:347-354.
5. AMCP Dossier for BrineuraTM (cerliponase alfa), BioMarin Pharmaceutical, May 2017.
6. Schulz A, et al. Study of Intraventricular Cerliponase Alfa for CLN2 Disease. N Engl J Med. 2018 Apr 24.