

Clinical Criteria

Subject:	Brineura (cerliponase alfa)	Publish Date:	04/01/2019 10/14/2019
Document #:	ING-CC-0012	Last Review Date:	08/17/2018 09/09/2019
Status:	Revised		

Formatted Table

Table of Contents

- [Overview](#)
- [Coding](#)
- [References](#)
- [Clinical criteria](#)
- [Document history](#)

Overview

This document addresses the use of Brineura (cerliponase alfa), a recombinant human tripeptidyl peptidase 1 enzyme replacement therapy for the treatment of late infantile neuronal ceroid lipofuscinosis type 2 (CLN2).

CLN2 is a form of Batten disease, a fatal inherited disorder of the nervous system, and results from a functional reduction in tripeptidyl peptidase 1 (TPP1) which is an enzyme that degrades proteins. Absence of TPP1 results in the accumulation of lysosomal storage materials in the brain and other organs and leads to progressive neurodegeneration and loss of cognitive, motor and visual function.

Brineura is approved to slow the loss of ambulation in symptomatic individuals with CLN2.

Clinical Criteria

When a drug is being reviewed for coverage under a member's medical benefit plan or is otherwise subject to clinical review (including prior authorization), the following criteria will be used to determine whether the drug meets any applicable medical necessity requirements for the intended/prescribed purpose.

Brineura (cerliponase alfa)

Requests for Brineura (cerliponase alfa) may be approved for:

- I. Symptomatic individuals with a diagnosis of late infantile neuronal ceroid lipofuscinosis type 2 (CLN2) as confirmed by:
 - A. Tripeptidyl peptidase 1 (TPP1) deficiency; **OR**
 - B. Detection of pathogenic mutations in each allele of the TPP1 gene (also known as the neuronal ceroid lipofuscinosis type 2 gene);

AND

II. Treatment is being given to slow the loss of ambulation.; **AND**

Brineura (cerliponase alfa) may not be approved for the following:

- II. There ~~are is no documentation of~~ acute intraventricular access device-related complications (such as leakage, device failure, or device-related infection) or ventriculoperitoneal shunts; **OR**
- II. Individual has signs or symptoms of acute or unresolved localized infection on or around the device insertion site (such as, cellulitis or abscess); **OR**
- III. Individual has suspected or confirmed central nervous system (CNS) infection (such as, cloudy cerebrospinal fluid [CSF], or positive CSF gram stain, or meningitis).

Requests for Brineura (cerliponase alfa) may not be approved when the above criteria are not met and for all other indications.

Formatted: Font: Not Bold

Formatted: Normal, Indent: Left: 0.5", No bullets or numbering

Formatted: No underline

Formatted: Font: (Default) Arial, 9 pt

Formatted: Normal, No bullets or numbering

Formatted: Numbered + Level: 1 + Numbering Style: I, II, III, ... + Start at: 1 + Alignment: Right + Aligned at: 0.19" + Indent at: 0.44"

Formatted: Font: Bold

Formatted: No underline

Formatted: No underline

Formatted: Indent: Left: 0.44", No bullets or numbering

Formatted: Right

Coding

The following codes for treatments and procedures applicable to this document are included below for informational purposes. Inclusion or exclusion of a procedure, diagnosis or device code(s) does not constitute or imply member coverage or provider reimbursement policy. Please refer to the member's contract benefits in effect at the time of service to determine coverage or non-coverage of these services as it applies to an individual member.

HCPCS

J0567 Injection, cerliponase alfa, 1 mg [Brineura]

Formatted: Font: Bold

ICD-10 Diagnosis

E75.4 Neuronal ceroid lipofuscinosis

Formatted: Font: Bold

Document History

Revised: 09/09/2019

Document History:

- 09/09/2019 – Annual Review: Update criteria to add labeled contraindication in those with localized infection around insertion site and those with CNS infections. Minor wording and formatting changes. Coding reviewed: No changes.
- 11/09/2018 – Coding Review: no changes.
- 11/21/2018 – Delete HCPCS codes: J3490 and C9014. Add code J0567.
- 08/17/2018 – Annual Review: Initial review of DRUG.00099. Minor wording and formatting changes.

References

1. Clinical Pharmacology [database online]. Tampa, FL: Gold Standard, Inc.; 2019. URL: <http://www.clinicalpharmacology.com>. Updated periodically.
2. DailyMed. Package inserts. U.S. National Library of Medicine, National Institutes of Health website. <http://dailymed.nlm.nih.gov/dailymed/about.cfm>. Accessed: August 26, 2019.
3. DrugPoints® System [electronic version]. Truven Health Analytics, Greenwood Village, CO. Updated periodically.
4. Lexi-Comp ONLINE™ with AHFS™. Hudson, Ohio: Lexi-Comp, Inc.; 2019; Updated periodically.
5. Online Mendelian Inheritance in Man (OMIM) #204500 Ceroid Lipofuscinosis, Neuronal, 2; CLN2. Available from: <https://www.omim.org/entry/204500>. Accessed on: August 26, 2019.

Federal and state laws or requirements, contract language, and Plan utilization management programs or polices may take precedence over the application of this clinical criteria.

No part of this publication may be reproduced, stored in a retrieval system or transmitted, in any form or by any means, electronic, mechanical, photocopying, or otherwise, without permission from the health plan.

© CPT Only – American Medical Association

Formatted: Right