Brineura™ (cerliponase alfa)

Last Review Date: November 1, 2019  Number: MG.MM.PH.41

Medical Guideline Disclaimer

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Definitions

Brineura is a hydrolytic lysosomal N-terminal tripeptidyl peptidase 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. Brineura is supplied as an injection for intraventricular administration.

Dosing and Administration

Brineura Package Insert

Guideline

Brineura is considered medically necessary for members ≥ 3 years of age with late infantile neuronal ceroid lipofuscinosis type 2 (CLN2) (also known as tripeptidyl peptidase 1 [TPP1] deficiency), when all of the following criteria are met:

1. Definitive diagnosis of late infantile CLN2 confirmed by TPP1 deficiency or the detection of pathogenic mutations in each allele of the TPP1 gene (aka CLN2 gene)

2. Presence of mild to moderate disease documented by a two-domain score of 3–6 on motor and language domains of the Hamburg CLN2 Clinical Rating Scale, with a score of at least 1 in each of these two domains

3. Member is ambulatory

4. Member does not have ventriculoperitoneal shunts
5. Member does not have acute intraventricular access device-related complications (e.g., leakage, device failure, or device-related infection)

Authorization

Initial authorization period of 6 months

Renewal based upon all:

1. Member continues to meet the criteria above
2. Absence of unacceptable toxicity from the drug or complications from the device (e.g., intraventricular access device leakage or infection, severe hypersensitivity reaction, severe hypotension; etc.)
3. 12-lead ECG evaluation performed within the last 6 months (those with cardiac abnormalities require ECG during each infusion)
4. Positive response to therapy compared to pretreatment baseline with stability/lack of decline in motor function/milestones on the Motor domain of the Hamburg CLN2 Clinical Rating Scale (decline is defined as having an unreversed [sustained] 2-category decline or an unreversed score of 0)

Limitations/Exclusions

Brineura is considered investigational when used for any indication not listed above.

Revisions

12/3/2018 Added J0567 and removed J3590 from Applicable Procedure Codes.

Applicable Procedure Codes

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<tr>
<th>Code</th>
<th>Description</th>
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<td>J0567</td>
<td>Injection, cerliponase alfa, 1 mg</td>
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Applicable Diagnosis Codes

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<th>Code</th>
<th>Description</th>
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<tr>
<td>E75.4</td>
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References


5. Specialty matched clinical peer review.