Crysvita® (burosumab-twza)

Last Review Date: October 14, 2019  Number: MG.MM.PH.116

Medical Guideline Disclaimer

Property of EmblemHealth. All rights reserved. The treating physician or primary care provider must submit to EmblemHealth the clinical evidence that the patient meets the criteria for the treatment or surgical procedure. Without this documentation and information, EmblemHealth will not be able to properly review the request for prior authorization. The clinical review criteria expressed below reflects how EmblemHealth determines whether certain services or supplies are medically necessary. EmblemHealth established the clinical review criteria based upon a review of currently available clinical information (including clinical outcome studies in the peer-reviewed published medical literature, regulatory status of the technology, evidence-based guidelines of public health and health research agencies, evidence-based guidelines and positions of leading national health professional organizations, views of physicians practicing in relevant clinical areas, and other relevant factors). EmblemHealth expressly reserves the right to revise these conclusions as clinical information changes, and welcomes further relevant information. Each benefit program defines which services are covered. The conclusion that a particular service or supply is medically necessary does not constitute a representation or warranty that this service or supply is covered and/or paid for by EmblemHealth, as some programs exclude coverage for services or supplies that EmblemHealth considers medically necessary. If there is a discrepancy between this guideline and a member’s benefits program, the benefits program will govern. In addition, coverage may be mandated by applicable legal requirements of a state, the Federal Government or the Centers for Medicare & Medicaid Services (CMS) for Medicare and Medicaid members. All coding and web site links are accurate at time of publication. EmblemHealth Services Company LLC, (“EmblemHealth”) has adopted the herein policy in providing management, administrative and other services to HIP Health Plan of New York, HIP Insurance Company of New York, Group Health Incorporated, GHI HMO Select, ConnectiCare, Inc., ConnectiCare Insurance Company, Inc. ConnectiCare Benefits, Inc., and ConnectiCare of Massachusetts, Inc. related to health benefit plans offered by these entities. All of the aforementioned entities are affiliated companies under common control of EmblemHealth Inc.

Definitions

Crysvita® (burosumab-twza) is a human immunoglobulin G subclass 1 (IgG1), anti-human fibroblast growth factor 23 (FGF23) antibody that binds to FGF23. FGF23 limits the reabsorption of phosphate in the renal tubules, which reduce the production of 1, 25-dihydroxy vitamin D in the kidneys. Burosumab-twza is indicated to treat patients > 1 year of age who have X-linked hypophosphatemia (XLH).¹

Dosing and Administration

For subcutaneous use only.

• Pediatric XLH:
  - For patients who weigh less than 10 kg, starting dose regimen is 1 mg/kg of body weight rounded to the nearest 1 mg, administered every two weeks
  - For patients who weigh more than 10 kg, starting dose regimen is 0.8 mg/kg of body weight rounded to the nearest 10 mg, administered every two weeks. The minimum starting dose is 10 mg up to a maximum dose of 90 mg.
  - Dose may be increased up to approximately 2 mg/kg (maximum 90 mg), administered every two weeks to achieve normal serum phosphorus.
• **Adult XLH:** Dose regimen is 1 mg/kg body weight rounded to the nearest 10 mg up to a maximum dose of 90 mg administered every four weeks.

**Initial Guideline**

Initial coverage is provided for **6 months** when all of the following criteria are met:

- Patient is at least 1 year of age; AND
- Patient has not received oral phosphate and/or active vitamin D analogs within 1 week prior to the start of therapy; AND
- Must be prescribed by, or in consultation with, a nephrologist or endocrinologist; AND
- Patient has a diagnosis of X-linked Hypophosphatemia (XLH) which is confirmed by at least one of the following:
  - Serum fibroblast growth factor-23 (FGF23) level > 30 pg/mL; OR
  - Phosphate regulating gene with homology to endopeptidases located on the X chromosome (PHEX-gene) mutations in the patient; AND
  - Patient has a reduced tubular resorption of phosphate corrected for glomerular filtration rate (TmP/GFR); AND
- Baseline fasting serum phosphorus* level with current hypophosphatemia, defined as a phosphate level below the lower limit of the laboratory normal reference range; AND
- Patient does not have severe renal impairment, defined as a glomerular filtration rate (GFR) of <30 mL/min; AND
- Patient presents with clinical signs and symptoms of the disease (e.g., rickets, growth retardation, musculoskeletal pain, bone fractures )

*Note: Phosphorous levels should be obtained fasting 12 hours or more without food or drink except for water and after an adequate washout period after supplements; lab values (i.e. GFR, phosphorous, TmP/GFR) should be obtained within 28 days of the date of administration.

**Renewal Guideline**

Coverage will be renewed for **12 months** when all of the following criteria are met:

- Patient continues to meet the criteria identified in the Initial Guideline section; AND
- Absence of unacceptable toxicity from the drug. Examples of unacceptable toxicity include the following: severe hypersensitivity reactions, hyperphosphatemia and/or nephrocalcinosis, severe injection site reactions, etc.; AND
• Patient has experienced normalization of serum phosphate within normal limits while on therapy; AND

• Disease response as indicated by increased serum phosphorus levels, a reduction in serum total alkaline phosphatase activity, improvement in symptoms (e.g., skeletal pain, linear growth, reduction of fractures, etc.), and/or improvement in radiographic imaging of Rickets/osteomalacia

Revisions

<table>
<thead>
<tr>
<th>Date</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>10/14/2019</td>
<td>Under Pediatric XLH, updated dosing guidelines to match PI (weight based)</td>
</tr>
<tr>
<td>12/03/2018</td>
<td>Added J0584 and removed J3590 from Applicable Procedure Codes</td>
</tr>
</tbody>
</table>

Applicable Procedure Codes

<table>
<thead>
<tr>
<th>Code</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>J0584</td>
<td>Injection, burosumab-twza, 1 mg</td>
</tr>
</tbody>
</table>

Applicable Diagnosis Codes

<table>
<thead>
<tr>
<th>Code</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>E83.30</td>
<td>Disorder of phosphorus metabolism, unspecified</td>
</tr>
<tr>
<td>E83.31</td>
<td>Familial hypophosphatemia</td>
</tr>
</tbody>
</table>

References

1 Crysvita [Package Insert]. Novato, CA; Ultragenyx Pharmaceutical; September 2019.