Medical Coverage Policy | Crysvita (burosumab-twza)

EFFECTIVE DATE: 08|01|2019
POLICY LAST UPDATED: 04|16|2019

OVERVIEW

Crysvita™ (burosumab-twza) is for the reatment of X-linked hypophosphatemia (XLH) in adult and pediatric patients 1 year of age and older.

This policy is applicable to BlueCHiP for Medicare products only. For Commercial Products, see related policy section.

MEDICAL CRITERIA

Initial Evaluation

Crysvita™ (burosumab-twza) will be approved when ALL of the following are met:

1. There is documentation that the patient is currently being treated with the requested agent

OR

2. All of the following:
    a. The patient has a diagnosis of X-linked hypophosphatemia (XLH) as confirmed by ONE of the following:
        i. Renal phosphate wasting AND radiography
        OR
        ii. Genetic testing AND fibroblast growth factor 23 (FGF23) level that is higher than the upper limit of normal

    AND

    b. The patient is 1 year of age or greater

    AND

    c. ONE of the following:
        i. The patient's epiphyseal plate has not fused
        OR
        ii. The patient's epiphyseal plate has fused AND the patient is experiencing symptoms of XLH (e.g. fractures, limited mobility)

    AND

    d. ONE of the following:
        i. The patient has tried and had an inadequate response to therapy with calcitriol and phosphate supplementation
        OR
        ii. The patient has a documented intolerance, FDA labeled contraindication, or hypersensitivity to calcitriol or phosphate supplementation
        OR
        iii. The prescriber has submitted documentation in support of therapy with the requested agent instead of calcitriol and phosphate supplementation therapy

    AND

    e. The prescriber is a specialist or has consulted with a specialist (e.g. endocrinologist, nephrologist)

    AND

    f. The patient does NOT have an FDA labeled contraindication to the requested agent

    AND

    g. The requested dosing is within FDA labeled dosing

Length of Approval: 12 months

Renewal Evaluation
Crysvita™ (burosumab-twza) will be approved when ALL of the following are met:

1. The patient has been previously approved for the requested agent through the Prime Therapeutics or BCBSRI PA or Medical Review process

AND

2. The patient has received clinical benefit from the requested agent (e.g. stabilization or slowing of disease progression, decrease in symptom severity and/or frequency)

AND

3. ONE of the following:
    a. The requested dose is within FDA labeling

    OR

    b. The requested dose for the requested diagnosis is supported by compendia (NCCN Compendium™[level of evidence 1, 2A], AHFS, DrugDex [FDA approved Class I or Class IIa])

    OR

    c. The requested dose is outside the dose supported by FDA labeling or compendia AND the patient is currently taking and is stable on this dose

    OR

    d. The request is for a change in dose that is outside that supported by FDA labeling or compendia AND the prescriber has submitted documentation (the dose is supported by clinical research in 2 or more peer reviewed medical journals) in support of therapy with a higher dose for the requested diagnosis

Length of Approval: 12 months

PRIOR AUTHORIZATION
Prior authorization is required for BlueCHiP for Medicare.

POLICY STATEMENT
BlueCHiP for Medicare
Crysvita™ (burosumab-twza) is medically necessary when the criteria above have been met.

COVERAGE
Benefits may vary between groups and contracts. Please refer to the appropriate Benefit Booklet, Evidence of Coverage or Subscriber Agreement for applicable physician administered drug medically necessary benefits/coverage.

BACKGROUND
X-linked hypophosphatemia (XLH) is a disorder of renal phosphate wasting. It is the most common form of heritable rickets. XLH is often misdiagnosed as nutritional rickets, metaphyseal dysplasia, and physiologic bowing. Patients with XLH have normal serum levels of calcium, normal-to-high parathyroid hormone levels, increased (or normal) alkaline phosphatase activity, normal plasma calcidiol concentration, and normal or slightly reduced plasma calcitriol concentration. Radiographic images should exclude diagnoses of physiologic bowing and most skeletal dysplasias. Definite evidence of renal phosphate wasting is critical to obtain before initiating therapy. Urinary phosphate excretion above 100 mg/day from a 24-hour urine collection or a fractional excretion of filtered phosphate (FEPO4) above 5 percent from a random urine specimen is indicative of renal phosphate wasting in patients with hypophosphatemia.

The gene responsible for XLH was identified on chromosome Xp22. and named PHEX (Phosphate regulating Endopeptidase on the X chromosome). This gene codes for a cell surface-bound protein-cleaving enzyme.

PHEX is expressed predominantly in bone and teeth, and the altered function of this bone-derived endopeptidase causes the renal phenotypic abnormalities of XLH. A large number of inactivating mutations in PHEX can cause XLH and there is no obvious correlation between genotype and phenotype. In a study of 118 families with at least one case of hypophosphatemic rickets, PHEX mutations were found in 87 percent of familial cases and in 72 percent of sporadic cases.

The principal phosphatonin involved in the pathogenesis of XLH is fibroblast growth factor 23 (FGF23). Mutations in PHEX (in bone tissue) indirectly alter the degradation and production of FGF23, causing increased circulating levels of the phosphatonin. FGF23, in turn, acts as a counter-regulatory hormone to inhibit phosphate reabsorption by the sodium/phosphate cotransporter in the kidney, acting through specific FGF receptors with the important co-factor klotho protein. Elevated levels of FGF23 also appear to be an important common pathway for other forms of hereditary hypophosphatemic rickets, as well as tumor-induced osteomalacia, although the mechanisms for the increased FGF23 vary among these disorders.

CODING

BlueCHiP for Medicare

The following HCPCS code is covered when the medical criteria have been met:

J0584 Injection, burosumab-twza, 1 mg

RELATED POLICIES

Prior Authorization of Drugs

PUBLISHED

Provider Update, June 2019

REFERENCES

1. Crysvita prescribing information. Ultragenyx Pharm, Inc. April 2018.
2. Carpenter TO, Imel EA, et al. A clinician's guide to X-linked hypophosphatemia. Journal of Bone Mineral Research. 2011 July; 26(7): 1381-1388.
3. Hereditary hypophosphatemic rickets and tumor-induced osteomalcia. UptoDate. Last updated 9/2017. Accessed 4/25/18.