Burosumab-twza (Crysvita®)

Meets Primary Coverage Criteria Or Is Covered For Contracts Without Primary Coverage Criteria
Initiation of Burosumab-twza (Cryvista®) meets member benefit certificate primary coverage criteria that there be scientific evidence of effectiveness for:

1. The treatment of X-linked hypophosphatemia (XLH) in adult and pediatric patients 6 months of age and older when ALL of the following requirements are met:
    a. PHEX (phosphate regulating endopeptidase homolog X-linked) gene mutation –(laboratory documentation must be provided) AND
    b. Serum fibroblast growth factor 23 (FGF23) level > 30 pg/mL by Kainos assay (laboratory documentation must be provided)
    c. Member's fasting serum phosphorus level is below age-based normal level (see below) –(laboratory documentation must be provided). Burosumab cannot be initiated if the serum phosphorus is within or above the normal range for age.

Age-Based Normal Serum Phosphate Reference Intervals
Age mg/dL mmol/L.
1-3 yrs 3.8-6.5 1.25-2.10
4-11 yrs 3.7-5.6 1.20-1.80
12-15 yrs 2.9-5.4 0.95-1.75
>15 yrs 2.7-4.7 0.90-1.50

    d. 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. bone pain, fractures, pseudofractures, limited ambulation).
        iii. Imaging or radiographic documentation must be provided at time of request, **AND**
    e. Use will not be in combination with oral phosphate or active vitamin D analogs AND
    f. The patient does not have severe renal impairment or end stage renal disease defined as a glomerular filtration rate < 30mL/min **AND**
    g. g. Evidence of reduced renal tubular phosphate reabsorption of phosphate corrected for glomerular filtration rate (TmP/GFR).

2. The treatment of FGF23-related hypophosphatemia in tumor-induced osteomalacia (TIO) in adult and pediatric patients 2 years of age and older when ALL of the following requirements are met:
    a. Member has a phosphaturic mesenchymal tumor cannot be curatively resected or cannot be located AND
    b. Member's FGF23 level ≥ 100pg/mL (laboratory documentation must be provided) (NCT02304367, 2020) **AND**
    c. Member's fasting serum phosphorus is below the normal range for age (laboratory documentation must be provided) **AND**

d. Member's ratio of renal tubular maximum reabsorption rate of phosphate to glomerular filtration rate (TmP/GFR) < 2.5mg/dL (NCT02304367, 2020) **AND**
e. Use will not be in combination with oral phosphate or active Vitamin D analogs AND
f. Member does not have severe renal impairment or end stage renal disease defined as a glomerular filtration rate < 30mL/min

Dosign and Administration

- Pediatric XLH (6 months and older):
  - 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 10 kg and greater, 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.
- Pediatric TIO (2 years and older): Starting dose is 0.4 mg/kg of body weight rounded to the nearest 10 mg every 2 weeks. Dose may be increased up to 2 mg/kg not to exceed 180 mg, administered every two weeks.
- Adult TIO: Starting dose is 0.5 mg/kg every four weeks. Dose may be increased up to 2 mg mg/kg not to exceed 180 mg, administered every two weeks.

Approval duration: 6 months

Continuation of burosumab-twza (Crysvita®) meets primary coverage criteria when **ALL** of the following criteria are met:

1. Authorization/reauthorization has been previously approved by Arkansas Blue Cross Blue Shield for treatment of x-linked hypophosphatemia (XLH), **OR** the member has previously met all indication-specific criteria.
2. The patient has demonstrated a clinically meaningful response to treatment with burosumab –documentation from the medical record must be provided.
3. Use will not be in combination with oral phosphate or active vitamin D analogs

Approval duration: 6 months