MASSACHUSETTS

Blue Cross Blue Shield of Massachusetts is an Independent Licensee of the Blue Cross and Blue Shield Association

Pharmacy Medical Policy

Retail Pharmacy Prior Authorization Policy

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Policy Number: 049

BCBSA Reference Number: None

Related Policies

- Quality Care Dosing guidelines apply to the following medications and can be found in Medical Policy #**621A**

Policy

Commercial Members: Managed Care (HMO and POS), PPO, and Indemnity

Note: All requests for outpatient retail pharmacy for indications listed and not listed on the medical policy guidelines may be submitted to BCBSMA Clinical Pharmacy Operations by completing the Prior Authorization Form on the last page of this document. Physicians may also call BCBSMA Pharmacy Operations department at (800)366-7778 to request a prior authorization/formulary exception verbally. Patients must have pharmacy benefits under their subscriber certificates.

Please refer to the chart below for the formulary and step status of the medications affected by this policy.

Standard Formulary	
Drug	**Formulary Status**
Arikayce ® (amikacin)	PA Required
Bylvay ™ (odevixibat)	PA Required
Dojolvi ™ (triheptanoin)	PA Required
Firdapse ® (amifampridine)	PA Required
Isturisa ® (osilodrostat)	PA Required
Livmarli ™ (maralixibat)	PA Required

Orladeyo ™ (berotralstat)	PA Required
Oxervate ™ (cenegermin)	PA Required
Pyrukynd ® (mitapivat)	PA Required
Recorlev ® (levoketoconazole)	PA Required
Targretin ® **Gel** (bexarotene)	PA Required
Tarpeyo ™ (budesonide)	PA Required
Tavneos ™ (avacopan)	PA Required
Voxzogo ™ (vosoritide)	PA Required
Vyndaqel ® (tafamidis meglumine)	PA Required
Vyndamax ® (tafamidis)	PA Required
Zokinvy (lonafarnib**)**	PA Required

We may cover Arikayce® (**amikacin suspension**)** for the treatment of adults, who have limited or no alternative treatment options, for the treatment of *Mycobacterium avium* complex (MAC) lung disease when **all** of the following criteria are met:

- Diagnosis of *Mycobacterium avium* complex (MAC) lung disease
- Minimum of 6 consecutive months of a multidrug background regimen therapy
- The drug is prescribed by a board-certified or board eligible Pulmonologist, or an Infectious Disease Specialist

We may cover Bylvay™ (**odevixibat**)** for the treatment of pruritus in patients 3 months of age and older with progressive familial intrahepatic cholestasis (PFIC) when **all** of the following criteria are met:

- Age is greater than or equal to three (3) months, **AND**
- Confirmed diagnosis of PFIC with molecular genetic testing, **AND**
- Molecular genetic testing does not indicate PFIC type 2 with *ABCB11* variants encoding for nonfunction or absence of BSEP-3, **AND**
- Presence of moderate to severe pruritis, **AND**
- Drug-induced pruritus has been ruled out, **AND**
- No history of liver transplant, **AND**
- No history of biliary diversion surgery within the past 6 months, **AND**
- No clinical evidence of decompensated cirrhosis

Note: If approved the Prior Authorization will be granted for up to one (1) year.

We may cover Dojolvi™ (**triheptanoin**)** for the treatment of pediatric and adult patients with molecularly confirmed long-chain fatty acid oxidation disorders (LC-FAOD) when **all** of the following criteria are met:

- molecularly confirmed long-chain fatty acid oxidation disorders (LC-FAOD)

AND

- The drug is prescribed by a board-certified or board eligible Endocrinologist, or a board-certified or board eligible Geneticist,

AND

- Frequent severe major medical episodes of hypoglycemia, rhabdomyolysis, or exacerbation of cardiomyopathy requiring emergency room visits, acute care visits, or hospitalizations **OR** Severe susceptibility to hypoglycemia or recurrent symptomatic hypoglycemia requiring intervention

  **Requests based exclusively on the use of samples will not meet coverage criteria for exception. Additional clinical information demonstrating medical necessity of the desired medication must be submitted by the requesting prescriber for review.

We may cover Firdapse® (**amifampridine**)** for the treatment of adults for the treatment of Lambert-Eaton myasthenic syndrome (LEMS) when **all** of the following criteria are met:

- Diagnosis of Lambert-Eaton myasthenic syndrome (LEMS)

We may cover Isturisa® (**osilodrostat**)** for the treatment of adults for the treatment of Cushing's Disease when **all** of the following criteria are met:

- Age is equal to or greater than 18 years, **AND**
- Confirmed documented diagnosis of Cushing's disease (**NOT** Cushing's Syndrome), **AND**
- The drug is prescribed by a board-certified or board eligible endocrinologist, **AND**
- Documentation of failed pituitary surgery or contraindication to pituitary surgery, **AND**
- Prescriber attests to the monitoring of Cortisol levels during titration and maintenance phase to ensure appropriate dose and adequate clinical response, **AND**
- This medication is not FDA approved for the treatment of Cushing's Syndrome.

Note: If approved the Prior Authorization will be granted for up to one (1) year.

We may cover Livmarli ™ (**maralixibat**)** for the treatment of cholestatic pruritus in patients with Alagille syndrome (ALGS) when **all** of the following criteria are met:

- Age is equal to or greater than 1 year of age, **AND**
- Confirmed documented diagnosis of Alagille Syndrome (ALGS), **AND**
- Presence of moderate to severe pruritis, **AND**

- Does not have chronic diarrhea requiring ongoing intravenous fluid or nutritional intervention, **AND**
- No history of liver transplant, **AND**
- No history of surgical interruption of enterohepatic circulation (for example, partial external biliary diversion [PEBD] surgery), **AND**
- No clinical evidence of decompensated cirrhosis

We may cover Orladeyo™ (berotralstat**) f**or prophylaxis to prevent attacks of hereditary angioedema (HAE)

- Age is equal to or greater than 12 years,

AND

- Diagnosis of HAE confirmed by low C4 or C1 inhibitor antigenic or functional level below lower limit of normal,

AND

- The drug is prescribed by a board-certified or board eligible Allergy & Immunology, Geneticist, or a Physician which specializes in the treatment of hereditary angioedema (HAE)

AND

- History of at least one moderate to severe acute attack per month such as airway swelling, severe abdominal pain, facial swelling, nausea and vomiting, or painful facial distortion

**Requests based exclusively on the use of samples will not meet coverage criteria for exception. Additional clinical information demonstrating medical necessity of the desired medication must be submitted by the requesting prescriber for review.

We may cover Oxervate™ (**cenegermin**)** eye drops for the treatment of adults with neurotrophic keratitis when **all** of the following criteria are met:

- Diagnosis of neurotrophic keratitis
- The approval is given for Eight (8) weeks of treatment

Note: If approved the Prior Authorization will be granted for up to 8 weeks.

We may cover Pyrukynd ® (mitapivat**)** for the treatment of adults with pyruvate kinase (PK) deficiency when **all** of the following criteria are met:

- Diagnosis of pyruvate kinase (PK) deficiency.
- Patient has at least two mutant alleles in the PKLR gene, of which at least one is a missense mutation.

Note: If approved the Prior Authorization will be granted for up to one (1) year.

We may cover Recorlev ® (levoketoconazole) may be covered when **ALL** of the following criteria must be met:

- The patient has hypercortisolemia and confirmed Cushing's syndrome
  And
- The patient is ≥ 18 years old,
  And
- The patient has had surgery and it was not curative or surgery is contraindicated
  And
- Documentation of baseline urinary free cortisol
  And
- Documentation of baseline live enzyme function tests

We may cover Targretin® (**bexarotene) for the topical treatment of cutaneous lesions in patients with cutaneous T-cell lymphoma (CTCL) when **all** of the following criteria are met:

- Diagnosis of refractory or persistent cutaneous T-cell lymphoma (CTCL)
- The Member has Stage IA or IB
- Document previous other therapies or a clinical rational for not using other therapies.

We may cover Tarpeyo ™** (budesonide) for reducing proteinuria in adults with primary immunoglobulin A nephropathy (IgAN) when **all** of the following criteria are met:

- Age is equal to or greater than 18 years, AND
- Diagnosis of primary immunoglobulin A nephropathy (IgAN), AND
- Document urine protein-to-creatinine ratio (UPCR) ≥1.5 g/g.

We may cover Tavneos ™** (avacopan) for adult patients with severe active anti-neutrophil cytoplasmic autoantibody (ANCA)-associated vasculitis (granulomatosis with polyangiitis [GPA] and microscopic polyangiitis [MPA]) when **all** of the following criteria are met:

- Age is equal to or greater than 18 years, AND
- Diagnosis of granulomatosis with polyangiitis [GPA] or microscopic polyangiitis [MPA], AND
- Used with glucocorticoids as part of standard therapy

We may cover Voxzogo ™** (vosoritide) for pediatric patients with achondroplasia when **all** of the following criteria are met:

- Age is equal to or greater than 5 years, AND
- Diagnosis of achondroplasia, AND
- Confirmed open epiphyses

Note: If approved the Prior Authorization will be granted for up to one (1) year.

We may cover Vyndaqel® (**tafamidis meglumine**)** or **Vyndamax®**** (tafamidis) when the patient has met **all** of the below criteria:

- Patient has a confirmed Diagnosis of Wild-type ATTR Amyloidosis (ATTRwt) **OR** hereditary transthyretin-mediated amyloidosis (ATTR-CM)

AND

- Patient is being treated for cardiomyopathy

AND

- Age is equal to or greater than 18 years

We may cover Zokinvy ® (**lonafarnib**)** when the patient has met **all** of the below criteria:

- Confirmed diagnosis of one of the following
    - Hutchinson-Gilford progeria syndrome (HGPS)
    - Heterozygous LMNA mutation with progerin-like protein accumulation
    - Homozygous or compound heterozygous ZMPSTE24 mutations

AND

- Patient has a BSA of at least 0.39 m^2

AND

- Age is equal to or greater than 12 months of age

AND

- Requested dose is appropriate for patient's BSA

Note: If approved the Prior Authorization will be granted for up to one (1) year.

**Requests based exclusively on the use of samples will not meet coverage criteria for exception. Additional clinical information demonstrating medical necessity of the desired medication must be submitted by the requesting prescriber for review.

For non-formulary/non-covered medications, requests must meet criteria above and the member must have had a previous treatment failure with or a contraindication to two covered formulary alternatives when available.

We do not cover the medications listed above for other conditions not listed above.

CPT Codes / HCPCS Codes / ICD-9 Codes

The following codes are included below for informational purposes. Inclusion or exclusion of a code does not constitute or imply member coverage or provider reimbursement. Please refer to the member's contract benefits in effect at the time of service to determine coverage or non-coverage as it applies to an individual member.

Providers should report all services using the most up-to-date industry-standard procedure, revenue, and diagnosis codes, including modifiers where applicable.

CPT Codes

There is no specific CPT code for this service.

Individual Consideration

All our medical policies are written for the majority of people with a given condition. Each policy is based on medical science. For many of our medical policies, each individual's unique clinical circumstances may be considered in light of current scientific literature. Physicians may send relevant clinical information for individual patients for consideration to:

Blue Cross Blue Shield of Massachusetts
Pharmacy Operations Department
25 Technology Place
Hingham, MA 02043
Tel: 1-800-366-7778
Fax: 1-800-583-6289

Prior Authorization Information

Outpatient

For services described in this policy, see below for products where prior authorization **IS REQUIRED** if the procedure is performed **outpatient**.

	Outpatient
Commercial Managed Care (HMO and POS)	Prior authorization is **required**.
Commercial PPO and Indemnity	Prior authorization is **required**.

Policy History

Date	Action
7/2022	Updated to add Pyrukynd to the policy.
4/2022	Updated to move Rinvoq ® to Step policy 010 and to add Recorlev ®.
2/2022	Updated to add Tarpeyo ™, Tavneos ™, Rinvoq ® , and Voxzogo ™ to the policy.
1/2022	Updated to add Livmarli ™ to the policy.
10/2021	Updated to add Bylvay ™ to the policy.
4/2021	Updated to add Zokinvy ® to the policy.
2/2021	Updated to add Orladeyo ™ to the policy.
10/2020	Updated to add Dojolvi ™ to the policy.
10/2020	Updated to add Targretin Gel to the policy.
9/2020	Updated to add Isturisa ® to the policy.
10/2019	Updated to add Vyndaqel ® & Vyndamax ®
8/2019	Updated to add Firdapse ® to the policy.
7/2019	Updated to add Oxervate ™ to the policy.
4/2019	Implementation of a new policy with a new to market medication Arikayce ®

References

1. Arikayce ® [package insert]. Bridgewater, NJ: Insmed, Inc.: 9/2018.
2. Oxervate ™ [package insert]. Boston, MA: Dompé U.S. Inc.: 11/2018.
3. Firdapse ® [package insert]. Coral Gables, FL: Catalyst Pharmaceuticals, Inc.: 11/2018.
4. Vyndaqel ® & Vyndamax ® [package insert]. New York, NY: Pfizer, Inc.: 8/2019.
5. Isturisa ® [package insert]. Lebanon, NJ: Recordati Rare Disease, Inc.: 3/2020.
6. Targretin ® Gel [package insert]. Bridgewater, NJ: Valeant Pharmaceuticals: 10/2016.
7. Dojolvi ™ [package insert]. Novato, CA: Ultragenyx Pharmaceutical Inc.: 6/2020.
8. Orladeyo ™ [package insert]. Durham, NC: BioCryst Pharmaceuticals, Inc.: 12/2020.

9. Zokinvy ™ [package insert]. Palo Alto, CA: Eiger BioPharmaceuticals, Inc.: 2/2021.
10. Bylvay ™ [package insert]. Boston, MA: Albireo Pharma, Inc.: 7/2021.
11. Livmarli ™ [package insert]. Foster City, CA: Mirum Pharmaceuticals, Inc.: 10/2021.
12. Tarpeyo ™ [package insert]. Stockholm, Sweden: Calliditas Therapeutics AB.: 12/2021.
13. Tavneos ™ [package insert]. San Carlos, CA: ChemoCentryx, Inc.: 10/2021
14. Voxzogo ™ [package insert]. Novato, CA: BioMarin Pharmaceutical Inc.: 12/2021
15. Recorlev ® [package insert]. Chicago, IL: Xeris Pharmaceuticals, Inc.: 1/2022.

To request prior authorization using the Massachusetts Standard Form for Medication Prior Authorization Requests (eForm), click the link below:
http://www.bluecrossma.org/medical-policies/sites/g/files/csphws2091/files/acquiadam-assets/023%20E%20Form%20medication%20prior%20auth%20instruction%20prn.pdf