Pharmacy Medical Policy

Drug Management & Retail Pharmacy Prior Authorization Policy

Table of Contents

- Policy: Commercial
- Policy: Medicare
- Coding Information
- Policy History
- Information Pertaining to All Policies
- References
- Endnotes
- Forms

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.
**Prior Authorization Information**

☒ Prior Authorization  ☐ Step Therapy  ☒ Quality Care Dosing

| Pharmacy (Rx) or Medical (MED) benefit coverage | ☒ Rx  ☐ MED |

Policy applies to Commercial Members:  
- Managed Care (HMO and POS),  
- PPO and Indemnity  
- MEDEX with Rx plan  
- Managed Major Medical with Custom BCBSMA Formulary  
- Comprehensive Managed Major Medical with Custom BCBSMA Formulary  
- Managed Blue for Seniors with Custom BCBSMA Formulary

To request for coverage: Physicians may call, fax, or mail the attached form (Formulary Exception/Prior Authorization form) to the address below.

Blue Cross Blue Shield of Massachusetts
Pharmacy Operations Department  
25 Technology Place  
Hingham, MA 02043

Individual Consideration: Policy for requests that do not meet clinical criteria of this policy, see section labeled Individual Consideration

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

**Standard Formulary**

<table>
<thead>
<tr>
<th>Drug</th>
<th>Formulary Status</th>
</tr>
</thead>
<tbody>
<tr>
<td>Arikayce ® (amikacin)</td>
<td>PA Required</td>
</tr>
<tr>
<td>bexarotene gel</td>
<td>PA Required</td>
</tr>
<tr>
<td>Bylvay ™ (odevixibat)</td>
<td>PA Required</td>
</tr>
<tr>
<td>Daybue ™ (trofinetide)</td>
<td>PA Required</td>
</tr>
<tr>
<td>Dojolvi ™ (triheptanoin)</td>
<td>PA Required</td>
</tr>
<tr>
<td>Filspari ™ (sparsentan)</td>
<td>PA Required</td>
</tr>
<tr>
<td>Firdapse ® (amifampridine)</td>
<td>PA Required</td>
</tr>
<tr>
<td>Isturisa ® (osilodrostat)</td>
<td>PA Required</td>
</tr>
<tr>
<td>Joenja ® (leniolisib)</td>
<td>PA Required</td>
</tr>
<tr>
<td>Livmarli ™ (maralixibat)</td>
<td>PA Required</td>
</tr>
<tr>
<td>Orladeyo ™ (berotralstat)</td>
<td>PA Required</td>
</tr>
<tr>
<td>Oxervate ™ (cenegermin)</td>
<td>PA Required</td>
</tr>
<tr>
<td>Pyrukynd ® (mitapivat)</td>
<td>PA Required</td>
</tr>
<tr>
<td>Recorlev ® (levoketoconazole)</td>
<td>PA Required</td>
</tr>
<tr>
<td>Skyclarys ™ (omaveloxolone)</td>
<td>PA Required</td>
</tr>
<tr>
<td>Targretin ® Gel (bexarotene)</td>
<td>PA Required</td>
</tr>
<tr>
<td>Tarpeyo ™ (budesonide)</td>
<td>PA Required</td>
</tr>
<tr>
<td>Tavneos ™ (avacopan)</td>
<td>PA Required</td>
</tr>
</tbody>
</table>
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, **AND**
- Minimum of 6 consecutive months of a multidrug background regimen therapy, **AND**
- 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 pruritus, **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

We may cover Daybue™ (trofinetide) for the treatment of Rett syndrome when ALL of the following criteria are met:

- diagnosis of Rett syndrome (mutations in MECP2 are not universal), **AND**
- Age is equal to or greater than 2 years

**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 Filspari™ (sparsentan) for the treatment of 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 biopsy verified Primary Immunoglobulin A nephropathy (IgAN), AND
- The urine total protein to creatinine ratio (UPCR) ≥ 1.5 g/g – submit UPCR.

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

- Diagnosis of Lambert-Eaton myasthenic syndrome (LEMS), AND
- Age is equal to or greater than 6 years

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.

We may cover Joenja® (leniolisib) for activated phosphoinositide 3-kinase delta (PI3Kδ) syndrome (APDS) when ALL of the following criteria are met:

- Age is equal to or greater than 12 years, AND
- Documented APDS/PASLI-associated PIK3CD/PIK3R1 mutation without concurrent use of immunosuppressive medication

**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 3 months of age, AND
- Confirmed documented diagnosis of Alagille Syndrome (ALGS), AND
- Presence of moderate to severe pruritus, 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) for 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, AND
- 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, AND
- Patient has at least two mutant alleles in the PKLR gene, of which at least one is a missense mutation.

We may cover Recorlev® (levoketoconazole) 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

**Note:** If approved the Prior Authorization will be granted for up to one (1) year.
We may cover **Skyclarys™** (omaveloxolone) for the treatment of Friedreich’s ataxia when ALL of the following criteria must be met:

- The patient has a genetically confirmed diagnosis of Friedreich's Ataxia, **AND**
- The patient is ≥ 16 years old, **AND**
- The drug is prescribed by a board-certified or board eligible Neurologist.

We may cover **Targretin®** (bexarotene) OR **Bexarotene** Gel 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), **AND**
- The Member has Stage IA or IB, **AND**
- 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 **Vijoice™** (avacopan) for patients with severe manifestations of PIK3CA-Related Overgrowth Spectrum (PROS) when ALL of the following criteria are met:

- Age is equal to or greater than two (2) years, **AND**
- Diagnosis of PIK3CA-Related Overgrowth Spectrum (PROS), **AND**
- Patient requires systemic 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

**Note:** If approved the Prior Authorization will be granted for up to one (1) year.
We may cover Xifaxan® (rifaximin) when the patient has met ALL of the below criteria:

• Patient has a confirmed Diagnosis of travelers’ diarrhea (TD) caused by noninvasive strains of Escherichia Coli, AND
• Age is equal to or greater than 12 years.

OR

• Patient has a confirmed diagnosis of irritable bowel syndrome with Diarrhea (IBS-D) or Small Intestinal Bacterial Overgrowth (SIBO), AND
• Age is equal to or greater than 18 years.

OR

• Patient is trying to reduce the risk of overt hepatic encephalopathy (HE) recurrence, 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
  o Hutchinson-Gilford progeria syndrome (HGPS)
  o Heterozygous LMNA mutation with progerin-like protein accumulation
  o Homozygous or compound heterozygous ZMPSTE24 mutations

AND

• Patient has a BSA of at least 0.39 m², 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

**Policy History**

<table>
<thead>
<tr>
<th>Date</th>
<th>Action</th>
</tr>
</thead>
<tbody>
<tr>
<td>7/2023</td>
<td>Updated Age for Livmarli™ and clarified Xifaxan® coding and added Filspari™, Skyclarys™, Daybue™, and Joenja® to the policy.</td>
</tr>
<tr>
<td>4/2023</td>
<td>Updated age for Firdapse®</td>
</tr>
<tr>
<td>1/2023</td>
<td>Updated to add Vioce™ and Xifaxan to the policy.</td>
</tr>
<tr>
<td>8/2022</td>
<td>Updated to add Bexarotene Gel to the policy.</td>
</tr>
<tr>
<td>7/2022</td>
<td>Updated to add Pyrukynd to the policy.</td>
</tr>
<tr>
<td>4/2022</td>
<td>Updated to move Rinvoq® to Step policy 010 and to add Recorlev®.</td>
</tr>
<tr>
<td>2/2022</td>
<td>Updated to add Tarpeyo™, Tavneos™, Rinvoq®, and Voxzogo™ to the policy.</td>
</tr>
<tr>
<td>1/2022</td>
<td>Updated to add Livmarli™ to the policy.</td>
</tr>
<tr>
<td>10/2021</td>
<td>Updated to add Bylvay™ to the policy.</td>
</tr>
<tr>
<td>4/2021</td>
<td>Updated to add Zokinvy® to the policy.</td>
</tr>
<tr>
<td>2/2021</td>
<td>Updated to add Orladeyo™ to the policy.</td>
</tr>
<tr>
<td>10/2020</td>
<td>Updated to add Dojolvi™ to the policy.</td>
</tr>
<tr>
<td>10/2020</td>
<td>Updated to add Targetretin Gel to the policy.</td>
</tr>
<tr>
<td>9/2020</td>
<td>Updated to add Isturisa® to the policy.</td>
</tr>
<tr>
<td>10/2019</td>
<td>Updated to add Vyndaqel® &amp; Vyndamax®</td>
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<tr>
<td>8/2019</td>
<td>Updated to add Firdapse® to the policy.</td>
</tr>
<tr>
<td>7/2019</td>
<td>Updated to add Oxervate™ to the policy.</td>
</tr>
<tr>
<td>4/2019</td>
<td>Implementation of a new policy with a new to market medication Arikayce®</td>
</tr>
</tbody>
</table>

**References**


To request prior authorization using the Massachusetts Standard Form for Medication Prior Authorization Requests (eForm), click the link below: