



MASSACHUSETTS

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Pharmacy Medical Policy

Drug Management & 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.

Prior Authorization Information

<input checked="" type="checkbox"/> Prior Authorization <input type="checkbox"/> Step Therapy <input checked="" type="checkbox"/> Quality Care Dosing		Pharmacy Operations: Tel: 1-800-366-7778 Fax: 1-800-583-6289
		Policy last updated 1/2024
Pharmacy (Rx) or Medical (MED) benefit coverage	<input checked="" type="checkbox"/> Rx <input type="checkbox"/> MED	To request for coverage: Providers 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
Policy applies to Commercial Members: <ul style="list-style-type: none"> • 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 		

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
bexarotene gel	PA Required
Bylvay ™ (odevixibat)	PA Required
Daybue ™ (trofinetide)	PA Required
Dojolvi ™ (triheptanoin)	PA Required
Filspari ™ (sparsentan)	PA Required
Firdapse ® (amifampridine)	PA Required
Isturisa ® (osilodrostat)	PA Required
Joenja ® (leniolisib)	PA Required
Livmarli ™ (maralixibat)	PA Required
Litfulo ™ (ritlecitinib)	PA Required
Orladeyo ™ (berotralstat)	PA Required
Oxervate ™ (cenegermin)	PA Required
Pyrukynd ® (mitapivat)	PA Required
Recorlev ® (levoketoconazole)	PA Required
Skyclarys ™ (omaveloxolone)	PA Required
Sohonos ™ (palovarotene)	PA Required
Targetin ® Gel (bexarotene)	PA Required

Standard Formulary -continued	
Tarpeyo ™ (budesonide)	PA Required
Tavneos ™ (avacopan)	PA Required
Vijoice ™ (alpelisib)	PA Required
Vivjoa ™ (oteseconazole)	PA Required
Voxzogo ™ (vosoritide)	PA Required
Vyndaqel ® (tafamidis meglumine)	PA Required
Vyndamax ® (tafamidis)	PA Required
Xifaxan ® (rifaximin)	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, **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 when **ALL** of the following criteria are met:

- Age is greater than or equal to three (3) months, **AND**
- Confirmed diagnosis of progressive familial intrahepatic cholestasis (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

OR

- Age is greater than or equal to twelve (12) months, **AND**
- Confirmed diagnosis of Alagille syndrome (ALGS), **AND**
- For the treatment of cholestatic pruritus

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 ^{TM**} (trihexanoin) 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 ^{TM**} (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 (PI3K δ) 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 ^{TM**} (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 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 Litfulo ^{TM**} (ritlecitinib) for the treatment of severe alopecia areata when **ALL** of the following criteria are met:

- Age is equal to or greater than 12 years of age, **AND**
- Confirmed diagnosis of alopecia areata
- Documentation of $\geq 50\%$ scalp hair loss
- Note: If approved the Prior Authorization will be granted for up to one (1) year

We may cover Orladeyo TM (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 ^{TM**} (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

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 Sohonos™ (palovarotene) for the treatment of Fibrodysplasia Ossificans Progressiva (FOP) when **ALL** of the following criteria must be met:

- The patient has a genetically (ACVR1) confirmed diagnosis of Fibrodysplasia Ossificans Progressiva (aka Myositis Ossificans Progressiva or Stoneman disease), **AND**
- The patient is born male ≥ 10 years old **OR** born female ≥ 8 years old, **AND**
- The drug is prescribed by a specialist in rare connective tissue diseases.

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

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 rationale 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 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 Vivjoa ^{TM**} (oteseconazole) for patients with recurrent vulvovaginal candidiasis (RVVC) with a history of RVVC which are NOT of reproductive potential when **all** of the following criteria are met:

- Age is equal to or greater than 12 years, **AND**
- Diagnosis of recurrent vulvovaginal candidiasis (RVVC), **AND**
- Patient has a history of RVVC (≥3 acute VVC episodes within 12 months), **AND**
- Patient is not of reproductive potential (i.e., history of tubal ligation, salpingo-oophorectomy, hysterectomy, or postmenopausal)

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

We may cover Voxzogo ^{TM**} (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
 - 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 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

Our medical policies are written for most people with a given condition. Each policy is based on peer reviewed clinical evidence. We also take into consideration the needs of atypical patient populations and diagnoses.

If the coverage criteria outlined is unlikely to be clinically effective for the prescribed purpose, the health care provider may request an exception to cover the requested medication based on an individual's unique clinical circumstances. This is also referred to as "individual consideration" or an "exception request."

Some reasons why you may need us to make an exception include: therapeutic contraindications; history of adverse effects; expected to be ineffective or likely to cause harm (physical, mental, or adverse reaction).

To facilitate a thorough and prompt review of an exception request, we encourage the provider to include additional supporting clinical documentation with their request. This may include:

- Clinical notes or supporting clinical statements.
- The name and strength of formulary alternatives tried and failed (if alternatives were tried) and specifics regarding the treatment failure, if applicable.
- Clinical literature from reputable peer reviewed journals.
- References from nationally recognized and approved drug compendia such as American Hospital Formulary Service® Drug Information (AHFS-DI), Lexi-Drug, Clinical Pharmacology, Micromedex or Drugdex®; and
- References from consensus documents and/or nationally sanctioned guidelines.

Providers may call, fax or mail relevant clinical information, including clinical references for individual patient consideration, to:

Blue Cross Blue Shield of Massachusetts
Pharmacy Operations Department

25 Technology Place
 Hingham, MA 02043
 Phone: 1-800-366-7778
 Fax: 1-800-583-6289

We may also use prescription claims records to establish prior use of formulary alternatives or to show if step therapy criteria has been met. We will require the provider to share additional information when prescription claims data is either not available or the medication fill history fails to establish use of preferred formulary medications or that step therapy criteria has been met.

Policy History

Date	Action
1/2024	Updated to add Sohonos [™] to the policy.
10/2023	Updated to add Litfulo [™] to the policy.
9/2023	Updated IC to align with 118E MGL § 51A.
7/2023	Updated Age for Livmarli [™] and clarified Xifaxan [®] coding and added Filspari [™] , Skyclarys [™] , Daybue [™] , and Joenja [®] to the policy.
4/2023	Updated age for Firdapse [®] .
1/2023	Updated to add Vjioice [™] and Xifaxan to the policy.
8/2022	Updated to add Bexarotene Gel to the policy
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: Insmmed, 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.
16. Xifaxan [®] [package insert]. Bridgewater, NJ: Salix Pharmaceuticals.: 10/2020.
17. Litfulo [™] [package insert]. New York, NY: Pfizer, Inc.: 6/2023.
18. Sohonos [™] [package insert]. Cambridge, MA: Ipsen Biopharmaceuticals, Inc.: 8/2023.

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>