Pharmacy Medical Policy
Drugs for Cystic Fibrosis

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Policy Number: 408
BCBSA Reference Number: None

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. Physicians may also submit requests for retail pharmacy exceptions via the web using Express PAth which can be found on the BCBSMA provider website or directly on the web at https://provider.express-path.com. 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.

<table>
<thead>
<tr>
<th>Drug</th>
<th>Formulary Information</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Standard</td>
</tr>
<tr>
<td></td>
<td>Formulary Status</td>
</tr>
<tr>
<td>Kalydeco™ (ivacaftor)</td>
<td>PA Required</td>
</tr>
<tr>
<td>Orkambi™ (lumacaftor / ivacaftor)</td>
<td>PA Required</td>
</tr>
<tr>
<td>Symdeko™ (tezacaftor / ivacaftor)</td>
<td>PA Required</td>
</tr>
</tbody>
</table>

We may cover Kalydeco™ (ivacaftor) for the treatment of cystic fibrosis when all of the following criteria are met¹:
- Age 1 years of age or older

We may cover Orkambi™ (lumacaftor and ivacaftor) for the treatment of cystic fibrosis when all of the following criteria are met¹:
- Age 2 years of age or older
- Documentation of **TWO** copies of the F508del mutation in the CFTR gene as confirmed by an FDA-cleared cystic fibrosis mutation test
- Concurrent use of **Kalydeco™** must be discontinued.

**We may cover Symdeko™** (tezacaftor and ivacaftor) for the treatment of cystic fibrosis when **all** of the following criteria are met:
- Age 12 years of age or older, **AND**
- Documentation of Homozygous for the F508del mutation in the CFTR gene as confirmed by an FDA-cleared cystic fibrosis mutation test
**OR**
**AND**
- Concurrent use of **Kalydeco™** or **Orkambi™** must be discontinued.

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

**Other Information**
Blue Cross Blue Shield of Massachusetts (BCBSMA*) members (other than Medex®; Blue MedicareRx, Medicare Advantage plans that include prescription drug coverage) will be required to fill their prescriptions for the above medications at one of the providers in our retail specialty pharmacy network, see link below:

[Link to Specialty Pharmacy List](#)

**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

**Managed Care Authorization Instructions**
- Physicians may call BCBSMA Pharmacy Operations department to request a review for prior authorization for patients.
  Pharmacy Operations: (800) 366-7778
- Physicians may also fax or mail the attached form to the address above. The Formulary Exception/Prior Authorization form is included as part of this document for physicians to submit for patients.
- Physicians may also submit requests for retail pharmacy exceptions via the web using Express Path which can be found on the BCBSMA provider website or directly on the web at [https://provider.express-path.com](https://provider.express-path.com).

**PPO and Indemnity Authorization Instructions**
- Physicians may call BCBSMA Pharmacy Operations department to request a review for prior authorization for patients.
  Pharmacy Operations: (800) 366-7778
- Physicians may also fax or mail the attached form to the address above. The Formulary Exception/Prior Authorization form is included as part of this document for physicians to submit for patients.
• Physicians may also submit requests for retail pharmacy exceptions via the web using Express PAth which can be found on the BCBSMA provider website or directly on the web at https://provider.express-path.com.

Policy History

<table>
<thead>
<tr>
<th>Date</th>
<th>Action</th>
</tr>
</thead>
<tbody>
<tr>
<td>9/2018</td>
<td>Updated to include new age range for Orkambi™ &amp; Kalydeco™.</td>
</tr>
<tr>
<td>6/2018</td>
<td>Updated to include Symdeko™ and to add Specialty Pharmacy Link.</td>
</tr>
<tr>
<td>10/2017</td>
<td>Updated to change Walgreens Specialty Name.</td>
</tr>
<tr>
<td>7/2017</td>
<td>Updated to include additional genes and add AllCare to Specialty pharmacy list.</td>
</tr>
<tr>
<td>6/2017</td>
<td>Updated address for Pharmacy Operations.</td>
</tr>
<tr>
<td>11/2016</td>
<td>Updated to include new age indication for Orkambi™.</td>
</tr>
<tr>
<td>4/2016</td>
<td>Updated to include Orkambi™ &amp; add Walgreens Specialty.</td>
</tr>
<tr>
<td>4/2015</td>
<td>Updated for new FDA approved ages.</td>
</tr>
<tr>
<td>2/2015</td>
<td>Updated new gene types which were FDA approved.</td>
</tr>
<tr>
<td>4/2014</td>
<td>Updated new gene types which were FDA approved.</td>
</tr>
<tr>
<td>2/2014</td>
<td>Removal of Curascript from Specialty Pharmacy section.</td>
</tr>
<tr>
<td>1/2014</td>
<td>Updated to remove Blue Value.</td>
</tr>
<tr>
<td>1/2013</td>
<td>New Policy, effective 1/1/2013.</td>
</tr>
</tbody>
</table>

References

Endnotes
1. Based on BCBSA Technology Evaluation Center Specialty Pharmacy Combined Capacity (SPCC) Report #3-2012 Ivacaftor (Kalydeco™), reviewed March 2012.

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