Updated: 11/2019
DMMA Approved: 12/2019

Request for Prior Authorization for Biologic Response Modifiers for Cystic Fibrosis
Website Form – www.highmarkhealthoptions.com
Submit request via: Fax - 1-855-476-4158

All requests for Biologic Response Modifiers for Cystic Fibrosis require a Prior Authorization and will be screened for medical necessity and appropriateness using the criteria listed below.

Biologic Response Modifiers for Cystic Fibrosis Prior Authorization Criteria:

Biologic Response Modifiers for Cystic Fibrosis include Kalydeco (ivacaftor), Symdeko (tezacaftor/ivacaftor), and Orkambi (lumacaftor/ivacaftor), and Trikafta (elexacaftor, tezacaftor, ivacaftor). New products with this classification will require the same documentation.

Coverage may be provided with a diagnosis of Cystic Fibrosis and the following criteria is met:

- Member is at least 2 years of age for Orkambi, 6 months of age for Kalydeco, 6 years of age for Symdeko, and 12 years of age for Trikafta.
- Has a documented genetic mutation as noted in the package labeling
- Documentation of baseline lab tests:
  - AST and ALT
  - FEV1
- Had all potential drug interactions addressed by the prescriber (such as discontinuation of the interacting drug, dose reduction of the interacting drug, or counseling of the member of the risks associated with the use of both medications when they interact
- The requested dose and frequency is in accordance with FDA-approved labeling, nationally recognized compendia, and/or evidence-based practice guidelines
- **Initial Duration of Approval:** 3 months
- **Reauthorization Criteria**
  - Had all potential drug interactions addressed by the prescriber (such as discontinuation of the interacting drug, dose reduction of the interacting drug, or counseling of the member of the risks associated with the use of both medications when they interact
  - Continues to benefit from treatment based on the prescriber's assessment
  - Repeat AST/ALT are ≤ 5 times the upper normal limit
  - For Orkambi,Symdeko, and Trikafta: must not have repeat lab results with AST/ALT > 3 times the upper normal limit **AND** bilirubin > 2 times the upper normal limit. If only one of these is elevated, continued use is appropriate.
- **Reauthorization Duration of Approval:** 12 months

Coverage may be provided for any non-FDA labeled indication if it is determined that the use is a medically accepted indication supported by nationally recognized pharmacy compendia or peer-reviewed medical literature for treatment of the diagnosis(es) for which it is prescribed. These requests will be reviewed on a case by case basis to determine medical necessity.

List of *CFTR* Gene Mutations that Produce CFTR Protein and are Responsive to KALYDECO (Table 1)

E56K	*G178R*	*S549R*	*K1060T*	*G1244E*
P67L	*E193K*	*G551D*	*A1067T*	*S1251N*
R74W	*L206W*	*G551S*	*G1069R*	*S1255P*
D110E	*R347H*	*D579G*	*R1070Q*	*D1270N*
D110H	*R352Q*	*S945L*	*R1070W*	*G1349D*
R117C	*A455E*	*S977F*	*F1074L*	*E831X*
R117H	*S549N*	*F1052V*	*D1152H*	*711+3A→G*
2789+5G→A	*3272-26A→G*	*3849+10kbC→T*		

List of *CFTR* Gene Mutations that Produce CFTR Protein and are Responsive to SYMDEKO (Table 2)

711+3A→G	*A455E*	*D579G*	*E193K*	*K1060T*	*R117C*	S945L
2789+5G→A	*A1067T*	*D1152H*	*E831X*	*L206W*	*R347H*	S977F
3272-26A→G	*D110E*	*D1270N*	*F1052V*	*P67L*	*R352Q*	
3849+10kbC→T	*D110H*	*E56K*	*F1074L*	*R74W*	*R1070W*	

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BIOLOGIC RESPONSE MODIFIERS FOR CYSTIC FIBROSIS
PRIOR AUTHORIZATION FORM

Please complete and fax all requested information below including any progress notes, laboratory test results, or chart documentation as applicable to Highmark Health Options Pharmacy Services. **FAX:** (855) 476-4158
If needed, you may call to speak to a Pharmacy Services Representative.
PHONE: (844) 325-6253 Monday through Friday 8:30am to 5:00pm

PROVIDER INFORMATION

Requesting Provider:	NPI:
Provider Specialty:	Office Contact:
Office Address:	Office Phone:
	Office Fax:

MEMBER INFORMATION

Member Name:	DOB:
Health Options ID:	Member weight: _____pounds or _____kg

REQUESTED DRUG INFORMATION

Medication:	Strength:
Frequency:	Duration:
Is the member currently receiving requested medication? ☐ Yes ☐ No	Date Medication Initiated:

Is this medication being used for a chronic or long-term condition for which the medication may be necessary for the life of the patient? ☐ Yes ☐ No

Billing Information

This medication will be billed: ☐ at a pharmacy **OR**
☐ medically (if medically please provide a JCODE:_____

Place of Service: ☐ Hospital ☐ Provider's office ☐ Member's home ☐ Other

Place of Service Information

Name:	NPI:
Address:	Phone:

MEDICAL HISTORY (Complete for ALL requests)

Diagnosis:
☐ Cystic Fibrosis

- Is there a genetic mutation as noted in the package labeling? ☐ Yes, please indicate the mutation: _____ ☐ No

☐ Other: _____ ICD-10: _____

Have baseline AST, ALT, and FEV1 been completed? ☐ Yes ☐ No

Have all potential drug interactions been addressed by the prescriber (such as discontinuation of the interacting drug, dose reduction of the interacting drug, or counseling of the member of the risks associated with the use of both medications when they interact)? ☐ Yes ☐ No

REAUTHORIZATION

Has the member experienced benefit from treatment? ☐ Yes ☐ No

Have all potential drug interactions been addressed by the prescriber (such as discontinuation of the interacting drug, dose reduction of the interacting drug, or counseling of the member of the risks associated with the use of both medications when they interact)? ☐ Yes ☐ No

Is the AST or ALT ≤ 5 times the upper limit of normal? ☐ Yes ☐ No

Orkambi, Symdeko, and Trikafta:

- Is the AST or ALT > 3 times the upper limit of normal? ☐ Yes ☐ No
- Is the bilirubin > 2 times the upper limit of normal? ☐ Yes ☐ No

SUPPORTING INFORMATION or CLINICAL RATIONALE	
Prescribing Provider Signature	Date