UnitedHealthcare Community Plan

Clinical Pharmacy Program Guidelines for Kalydeco

Program	Prior Authorization
Medication	Kalydeco™ (ivacaftor)
Issue Date	6/2015
Pharmacy and Therapeutics Approval Date	8/2017
Effective Date	10/2017

1. Background:

Kalydeco (ivacaftor) is a cystic fibrosis transmembrane conductance regulator (CFTR) potentiator indicated for the treatment of cystic fibrosis (CF) in patients age 2 years and older who have who have one of the following mutations in the CFTR gene:

A1067T	*E193K*	*G1349D*	*R1070W*	*S1255P*
A455E	*E56K*	*G178R*	*R117C*	*S549N*
D110E	*F1052V*	*G551S*	*R117H*	*S549R*
D110H	*F1074L*	*K1060T*	*R347H*	*S945L*
D1152H	*G1069R*	*L206W*	*R352Q*	*S977F*
D1270N	*G551D*	*P67L*	*R74W*	
D579G	*G1244E*	*R1070Q*	*S1251N*	

If the patient's genotype is unknown, an FDA-cleared CF mutation test should be used to detect the presence of a CFTR mutation followed by verification with bi-directional sequencing when recommended by the mutation test instructions for use. Kalydeco is not effective in patients with CF who are homozygous for the *F508del* mutation in the *CFTR* gene.[1]

Members will be required to meet the coverage criteria below.

2. Coverage Criteria:

A. <u>Initial Authorization</u>

1. Kalydeco will be approved based upon **<u>all</u>** of the following criteria:

a. Diagnosis of cystic fibrosis (CF)

-AND-

b. Submission of laboratory results confirming that patient has **<u>one</u>** of the following mutations in the CFTR gene:

A1067T	*E193K*	*G1349D*	*R1070W*	*S1255P*
A455E	*E56K*	*G178R*	*R117C*	*S549N*
D110E	*F1052V*	*G551S*	*R117H*	*S549R*
D110H	*F1074L*	*K1060T*	*R347H*	*S945L*
D1152H	*G1069R*	*L206W*	*R352Q*	*S977F*
D1270N	*G551D*	*P67L*	*R74W*	
D579G	*G1244E*	*R1070Q*	*S1251N*	

-AND-

c. Prescribed by or in consultation with a specialist affiliated with a CF care center

Authorization will be issued for 12 months.

B. Reauthorization

1. Kalydeco will be approved based on **both** of the following criteria:

a. Provider attests that the patient has achieved a clinically meaningful response while on Kalydeco therapy to **one** of the following:

(1) Lung function as demonstrated by percent predicted expiratory volume in 1 second (ppFEV$_1$)
(2) Body mass index (BMI)
(3) Pulmonary exacerbations
(4) Quality of life as demonstrated by Cystic Fibrosis Questionnaire-Revised (CFQ-R) respiratory domain score

-AND-

b. Prescribed by or in consultation with specialist affiliated with a CF care center

Authorization will be issued for 12 months.

3. References:

1. Kalydeco [Package Insert]. Cambridge, MA: Vertex Pharmaceuticals, Inc.; May 2017.

Program	Prior Authorization - Kalydeco (ivacaftor)
Change Control	
6/2015	New Program

7/2016	Updated policy template. Aligning with Employer & Individual on clinical criteria but changed reauthorization duration from 24 to 12 months.
11/2016	Revised prescriber criterion
3/2017	Changed initial authorization duration to 12 months
8/2017	Added 23 additional CFTR mutations based on labeling change