CareSource Rxinnovations

PHARMACY POLICY STATEMENT
Marketplace

DRUG NAME	Kalydeco (ivacaftor)
BILLING CODE	Must use valid NDC
BENEFIT TYPE	Pharmacy
SITE OF SERVICE ALLOWED	Home
STATUS	Prior Authorization Required

Kalydeco (ivacaftor) is a is a cystic fibrosis transmembrane conductance regulator (CFTR) potentiator initially approved by the FDA in 2012. Cystic fibrosis is an autosomal recessive disease in which patients can have abnormal airways secretions, chronic endobronchial infection, and progressive airway obstruction.

Kalydeco (ivacaftor) will be considered for coverage when the following criteria are met:

Cystic Fibrosis

For **initial** authorization:

1. Member is at least 4 months of age; AND
2. Medication must be prescribed by or in consultation with a pulmonologist or an infectious disease specialist; AND
3. Member has a diagnosis of cystic fibrosis; AND
4. Member has had genetic testing documented in chart notes with one of the following mutations in the CFTR gene: 711+3A→G, F311del, I148T, R75Q, S589N, 2789+5G→A, F311L, I175V, R117C,S737F, 3272-26A→G, F508C, I807M, R117G, S945L, 3849+10kbC→T, F508C;S1251N, I1027T, R117H, S977F, A120T, F1052V, I1139V, R117L, S1159F, A234D, F1074L, K1060T, R117P, S1159P, A349V, G178E, L206W, R170H, S1251N, A455E, G178R, L320V, R347H, S1255P, A1067T, G194R, L967S, R347L, T338I, D110E, G314E, L997F, R352Q, T1053I, D110H, G551D, L1480P, R553Q, V232D, D192G, G551S, M152V, R668C, V562I, D579G, G576A, M952I, R792G, V754M, D924N, G970D, M952T, R933G, V1293G, D1152H, G1069R, P67L, R1070Q, W1282R, D1270N, G1244E, Q237E, R1070W, Y1014C, E56K, G1249R, Q237H, R1162L, Y1032C, E193K, G1349D, Q359R, R1283M, E822K, H939R, Q1291R, S549N, E831X, H1375P, R74W, S549R
5. **Dosage allowed/Quantity limit:**
   a) Adult and pediatric members 6 years of age or older: 150 mg every 12 hours (60 tablets per 30 days)
   b) Pediatrics under 6 years of age (56 packets per 28 days):

Age	Dosage Allowed	
Infants 4 to <6 months old and weighing ≥5 kg	25 mg granule packet every 12 hours	
Infants ≥6 months and <6 years old	Weight	Dosage Allowed
	5 to <7 kg	25 mg packet every 12 hours
	7 to <14 kg	50 mg packet every 12 hours
	≥14 kg	75 mg packet every 12 hours

If all the above requirements are met, the medication will be approved for 3 months.

For **reauthorization**:

1. Member must be in compliance with all other initial criteria; AND
2. Member's adherence to medication is confirmed by claims history; AND
3. Chart notes must show improvement or stabilized signs and symptoms of disease demonstrated by any of the following:
   a) Improved FEV1 and/or other lung function tests
   b) Improvement in sweat chloride
   c) Decrease in pulmonary exacerbations
   d) Decrease in pulmonary infections
   e) Increase in weight-gain
   f) Decrease in hospitalizations

If all the above requirements are met, the medication will be approved for an additional 12 months.

CareSource considers Kalydeco (ivacaftor) not medically necessary for the treatment of conditions that are not listed in this document. For any other indication, please refer to the Off-Label policy.

DATE	ACTION/DESCRIPTION
06/12/2017	New policy for Kalydeco created.
10/05/2018	New CFTD gene mutations added. Age coverage expanded (approved for 12 months old members and older).
05/16/2019	Age coverage expanded (approved for 6 months old members and older).
12/30/2020	Policy reviewed. New age limit expanded to 4 months of age (previously 6 months). List of approved mutations expanded. Added dosing chart for patients 6 years of age and younger. Reauthorization criteria updated to ask for evidence of disease stability or improvement.
04/27/2022	Policy transferred to new template. Amended reference section.

References:

1. Kalydeco [package insert]. Boston, MA: Vertex Pharmaceuticals Inc; December, 2020.
2. Kalydeco. Micromedex Solutions. Truven Health Analytics, Inc. Ann Arbor, MI. Available at: http://www.micromedexsolutions.com. Accessed March 6, 2017.
3. Ren CL, Morgan RL, Oermann C, et al. Cystic Fibrosis Foundation Pulmonary Guidelines. Use of Cystic Fibrosis Transmembrane Conductance Regulator Modulator Therapy in Patients with Cystic Fibrosis. *Ann Am Thorac Soc*. 2018;15(3):271-280. doi:10.1513/AnnalsATS.201707-539OT.

Effective date: 10/01/2022
Revised date: 04/27/2022