CareSource Rx Innovations

PHARMACY POLICY STATEMENT
Georgia Medicaid

DRUG NAME	Kalydeco (ivacaftor)
BENEFIT TYPE	Pharmacy
STATUS	Prior Authorization Required

Kalydeco (ivacaftor) is a is a cystic fibrosis transmembrane conductance regulator (CFTR) potentiator indicated for the treatment of cystic fibrosis (CF) in patients age 1 month and older who have at least one mutation in the CFTR gene that is responsive to ivacaftor based on clinical and/or in vitro assay data. It facilitates increased chloride transport by potentiating the channel open probability (or gating) of CFTR protein located at the cell surface. The CFTR protein is a chloride channel present at the surface of epithelial cells in multiple organs.
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 1 month 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:** See table below. Quantity limit: 60 tablets per 30 days or 56 packets per 28 days.

Age	Weight	Dosage
1 month to less than 2 months	3 kg or greater	One 5.8 mg packet every 12 hours
2 months to less than 4 months	3 kg or greater	One 13.4 mg packet every 12 hours

4 months to less than 6 months	5 kg or greater	One 25 mg packet every 12 hours
6 months to less than 6 years	5 kg to less than 7 kg	One 25 mg packet every 12 hours
	7 kg to less than 14 kg	One 50 mg packet every 12 hours
	14 kg or greater	One 75 mg packet every 12 hours
6 years and older	-	One 150 mg tablet every 12 hours

If all the above requirements are met, the medication will be approved for 6 months.

For **reauthorization**:

1. 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.
05/22/2023	Lowered age limit to 1 month to align with FDA approval; removed compliance with initial criteria and adherence in claims history from reauthorization criteria; added reference.
01/28/2025	Updated references; increased initial authorization length from 3 months to 6 months.

References:

1. Kalydeco [package insert]. Boston, MA: Vertex Pharmaceuticals Inc; 2023.
2. 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.

3. Farrell PM, White TB, Ren CL, et al. Diagnosis of Cystic Fibrosis: Consensus Guidelines from the Cystic Fibrosis Foundation [published correction appears in J Pediatr. 2017 May;184:243]. J Pediatr. 2017;181S:S4-S15.e1. doi:10.1016/j.jpeds.2016.09.064
4. Southern KW, Castellani C, Lammertyn E, et al. Standards of care for CFTR variant-specific therapy (including modulators) for people with cystic fibrosis. J Cyst Fibros. 2023;22(1):17-30. doi:10.1016/j.jcf.2022.10.002

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