Reference number(s)
1884-A

Specialty Guideline Management Kalydeco

Products Referenced by this Document

Drugs that are listed in the following table include both brand and generic and all dosage forms and strengths unless otherwise stated. Over-the-counter (OTC) products are not included unless otherwise stated.

Brand Name	Generic Name
Kalydeco	ivacaftor

Indications

The indications below including FDA-approved indications and compendial uses are considered a covered benefit provided that all the approval criteria are met and the member has no exclusions to the prescribed therapy.

FDA-Approved Indication[1]

Kalydeco is indicated for the treatment of cystic fibrosis (CF) in patients age 1 month and older who have at least one mutation in the cystic fibrosis transmembrane conductance regulator (CFTR) gene that is responsive to ivacaftor potentiation based on clinical and/or in vitro assay data.

If the patient's genotype is unknown, an FDA-cleared CF mutation test should be used to detect the presence of CFTR mutation followed by verification with bi-directional sequencing when recommended by the mutation test instructions for use.

All other indications are considered experimental/investigational and are not medically necessary.

Documentation

Submission of the following information is necessary to initiate the prior authorization review: For initial requests, genetic testing report confirming the presence of the appropriate CFTR gene mutation.

© 2025 CVS Caremark. All rights reserved.

This document contains confidential and proprietary information of CVS Caremark and cannot be reproduced, distributed or printed without written permission from CVS Caremark. This document contains prescription brand name drugs that are trademarks or registered trademarks of pharmaceutical manufacturers that are not affiliated with CVS Caremark.

Prescriber Specialties

This medication must be prescribed by or in consultation with a pulmonologist.

Coverage Criteria

Cystic fibrosis[1,2]

Authorization of 12 months may be granted for treatment of cystic fibrosis when all the following criteria are met:

- Genetic testing was conducted to detect a mutation in the CFTR gene.
- The member has one of the following mutations in the CFTR gene: A120T, A234D, A349V, A455E, A1067T, D110E, D110H, D192G, D579G, D924N, D1152H, D1270N, E56K, E193K, E822K, E831X, F311del, F311L, F508C, S1251N, F1052V, F1074L, G178E, G178R, G194R, G314E, G551D, G551S, G576A, G970D, G1069R, G1244E, G1249R, G1349D, H939R, H1375P, I148T, I175V, I807M, I1027T, I1139V, K1060T, L206W, L320V, L967S, L997F, L1480P, M152V, M952I, M952T, P67L, Q237E, Q237H, Q359R, Q1291R, R74W, R75Q, R117C, R117G, R117H, R117L, R117P, R170H, R347H, R347L, R352Q, R553Q, R668C, R792G, R933G, R1070Q, R1070W, R1162L, R1283M, S549N, S549R, S589N, S737F, S945L, S977F, S1159F, S1159P, S1251N, S1255P, T338I, T1053I, V232D, V562I, V754M, V1293G, W1282R, Y1014C, Y1032C, 711+3A→G, 2789+5G→A, 3272-26A→G, 3849+10kbC→T.
- The member is at least 1 month of age.

Continuation of Therapy

Authorization of 12 months may be granted for continued treatment in members requesting reauthorization for an indication listed in the coverage criteria section who are experiencing benefit from therapy as evidenced by disease stability or disease improvement (e.g., improvement in FEV1 from baseline).

Other

Kalydeco will not be used in combination with another CFTR modulator for the treatment of cystic fibrosis (e.g., Orkambi, Symdeko).

References

© 2025 CVS Caremark. All rights reserved.

This document contains confidential and proprietary information of CVS Caremark and cannot be reproduced, distributed or printed without written permission from CVS Caremark. This document contains prescription brand name drugs that are trademarks or registered trademarks of pharmaceutical manufacturers that are not affiliated with CVS Caremark.

1. Kalydeco [package insert]. Boston, MA: Vertex Pharmaceuticals Inc.; August 2023.
2. Mogayzel PJ, Naureckas ET, Robinson KA, et al. Cystic fibrosis pulmonary guidelines. chronic medications for maintenance of lung health. Am J Respir Crit Care Med. 2013;187:680-689 doi: 10.1542/peds.2015-1784.

© 2025 CVS Caremark. All rights reserved.

This document contains confidential and proprietary information of CVS Caremark and cannot be reproduced, distributed or printed without written permission from CVS Caremark. This document contains prescription brand name drugs that are trademarks or registered trademarks of pharmaceutical manufacturers that are not affiliated with CVS Caremark.