SPECIALTY GUIDELINE MANAGEMENT

KALYDECO (ivacaftor)

POLICY

I. 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
Kalydeco 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 one mutation in the CFTR gene that is responsive to ivacaftor potentiation based on clinical and/or in vitro assay data.

All other indications are considered experimental/investigational and are not a covered benefit.

II. CRITERIA FOR INITIAL APPROVAL

A. Cystic Fibrosis

Indefinite authorization may be granted for treatment of cystic fibrosis when all of the following criteria are met:

1. Genetic testing was conducted to detect a mutation in the *CFTR* gene.
2. The member has one of the following mutations in the *CFTR* gene: A455E, A1067T, D110E, D110H, D579G, D1152H, D1270N, E56K, E193K, F1052V, F1074L, G178R, G551D, G551S, G1069R, G1244E, G1349D, K1060T, L206W, P67L, R74W, R117C, R117H, R347H, R352Q, R1070Q, R1070W, S549N, S549R, S945L, S977F, S1251N, or S1255P.
3. The member is at least 2 years of age.
4. Kalydeco will not be used in combination with Orkambi.

III. CONTINUATION OF THERAPY

All members (including new members) requesting authorization for continuation of therapy must meet all initial authorization criteria.

IV. REFERENCES

1. Kalydeco [package insert]. Boston, MA: Vertex Pharmaceuticals Inc.; May 2017.
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.