AETNA BETTER HEALTH® Coverage Policy/Guideline					
Name:	Kalydeco			Page:	1 of 2
Effective Date:	9/29/2023			Last Review Date:	8/11/2023
Applies to:	☒Illinois ☐New Jersey ☒Pennsylvania Kids	☐Florida ☒Maryland ☒Virginia		☒Florida Kids ☐Michigan ☐Kentucky PRMD	

Intent:

The intent of this policy/guideline is to provide information to the prescribing practitioner outlining the coverage criteria for Kalydeco under the patient's prescription drug benefit.

Description:

FDA-Approved Indication
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.

Applicable Drug List:

Kalydeco

Policy/Guideline:

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

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

Criteria for Initial Approval:
Cystic Fibrosis
Authorization of 12 months may be granted for treatment of cystic fibrosis when all of the following criteria are met:

A. Genetic testing was conducted to detect a mutation in the *CFTR* gene.
B. 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, 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.

C. The member is at least 4 months of age.
D. Kalydeco will not be used in combination with other medications containing ivacaftor.

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

Approval Duration and Quantity Restrictions:
Approval: 12 months

Quantity Level Limit:

- Kalydeco (ivacaftor) tablets 150 mg blister carton: 1 carton (56 tablets) per 28 days
- Kalydeco (ivacaftor) tablets 150 mg: 60 tablets per 30 days
- Kalydeco (ivacaftor) oral granules 5.8 mg packets: 56 packets per 28 days
- Kalydeco (ivacaftor) oral granules 13.4 mg packets: 56 packets per 28 days
- Kalydeco (ivacaftor) oral granules 25 mg packets: 56 packets per 28 days
- Kalydeco (ivacaftor) oral granules 50 mg packets: 56 packets per 28 days
- Kalydeco (ivacaftor) oral granules 75 mg packets: 56 packets per 28 days

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