Kalydeco (ivacaftor)

Override(s)	Approval Duration
Prior Authorization Quantity Limit	1 year

Medications	Quantity Limit
Kalydeco (ivacaftor)	May be subject to quantity limit

APPROVAL CRITERIA

Initial requests for Kalydeco (ivacaftor) may be approved if the following are met:

I. Individual has a diagnosis of cystic fibrosis (CF); **AND**
II. Individual is 1 month of age or older; **AND**
III. A copy of the CF mutation analysis test results must be provided; **AND**
IV. Individual has a mutation-positive result in the cystic fibrosis transmembrane conductance regulator (CFTR) gene with **one** of the following mutation types:

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	

Continuation requests for Kalydeco (ivacaftor) may be approved if the following is met:

I. There is clinically significant improvement or stabilization in clinical signs and symptoms of disease (including but not limited to improvement in FEV1, decrease in pulmonary

exacerbations, improvement in BMI or improvement of respiratory symptoms [cough, sputum production, difficulty breathing]).

Kalydeco (ivacaftor) monotherapy, may not be approved for the following:

I. Individual is homozygous for F508del mutation in the CFTR gene; **OR**
II. Individual with hepatic impairment if less than 6 months of age; **OR**
III. In combination with Orkambi (ivacaftor/lumacaftor), Symdeko (tezacaftor/ivacaftor) or Trikafta (elexacaftor/tezacaftor/ivacaftor).

Key References:

1. DailyMed. Package inserts. U.S. National Library of Medicine, National Institutes of Health website. http://dailymed.nlm.nih.gov/dailymed/about.cfm. Accessed: March 19, 2023.
2. DrugPoints® System [electronic version]. Truven Health Analytics, Greenwood Village, CO. Updated periodically.
3. Lexi-Comp ONLINE™ with AHFS™, Hudson, Ohio: Lexi-Comp, Inc. Updated periodically.
4. Simon RH. Cystic fibrosis: Overview of the treatment of lung disease. Last updated: February 13, 2023. In: UpToDate, Post TW (Ed), UpToDate, Waltham, MA. Accessed: March 17, 2023.
5. Simon RH. Cystic fibrosis: Treatment with CFTR modulators. Last updated: November 29, 2022. In: UpToDate, Post TW (Ed), UpToDate, Waltham, MA. Accessed: March 17, 2023.