



Prior Authorization Criteria

Kalydeco® (*ivacaftor*) PA CRITERIA:

Kalydeco is indicated for the treatment of cystic fibrosis in patients age 4 months 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.

Select the diagnosis:

Cystic fibrosis (CF) ICD-10 code(s): _____

Initial authorization: 6 months

Prior authorization approval will be considered when **ALL** of the following criteria are met:

Yes No Age of patient is within the age range as recommended by the FDA label* **AND**

Yes No Prescribed by or in consultation with a CF specialist/ pulmonologist who specializes in treating CF patients; **AND**

a. Name of CF treating/consulting specialist/pulmonologist

b. For consults, provide chart documentation including name of drug

Yes No Patient has a diagnosis of cystic fibrosis (CF) and has *one* CFTR mutation responsive to Kalydeco** 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 a CFTR mutation followed by verification with bi-directional sequencing when recommended by the mutation test instructions for use. Submission, upon request, of laboratory results documenting responsive CFTR mutation; **AND**

**CFTR Mutations Responsive to Kalydeco

(Continued next page)

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*	

* Clinical data exist for these mutations [see Clinical Studies (14)].

† Complex/compound mutations where a single allele of the CFTR gene has multiple mutations; these exist independent of the presence of mutations on the other allele.

F508del and 26 other mutations are considered not responsive to ivacaftor (see Prescribing Information for complete listing).

- Yes No Baseline measures submitted by provider of ALL of the following:
- For age appropriate patients, percent predicted expiratory volume in 1 second (ppFEV1): _____
 - Body mass index (BMI): _____
 - Pulmonary exacerbations- number in preceding 6 months: _____

Reauthorization: 12 months with evidence of appropriate clinical response to therapy

Yes No Prescribed by or in consultation with a CF specialist/ pulmonologist who specializes in treating CF patients.

- Name of CF treating/consulting specialist/pulmonologist

- For consults, provide chart documentation including name of drug

AND

Yes No Provider attests that the patient has achieved a clinically meaningful response while on Kalydeco based on ALL of the following:

- For age appropriate patients, improved or stable lung function as demonstrated by percent predicted expiratory volume in 1 second (ppFEV1): _____
- Body mass index (BMI): _____

- c. Pulmonary exacerbations- number of exacerbations compared to number of exacerbations prior to medication initiation: _____

How Supplied:

Kalydeco (ivacaftor) tablets

60-count bottle 150 mg tablets

56-count carton (contains 4 individual blister cards of 14 tablets per card)

Kalydeco (ivacaftor) oral granules (for use in children age less than 6 years)

- ****Use of granules for children equal to or greater than 6 years requires clinical justification***
 - 56-count carton (contains 56 unit-dose packets of 25mg ivacaftor per packet)
 - 56-count carton (contains 56 unit-dose packets of 50mg ivacaftor per packet)
 - 56-count carton (contains 56 unit-dose packets of 75 mg ivacaftor per packet)