Prior Authorization Criteria

MISSISSIPPI DIVISION OF MEDICAID

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:

o 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:

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

o Yes o 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
_____

o Yes o 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 CTFR mutation; **AND**

**CFTR Mutations Responsive to Kalydeco

(Continued next page)

KALYDECO® (ivacaftor) Tablets and Oral Granules

Table 3: List of *CFTR* Gene Mutations that Produce CFTR Protein and are Responsive to KALYDECO				
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).

o Yes o No Baseline measures submitted by provider of ALL of the following:

a. For age appropriate patients, percent predicted expiratory volume in 1 second (ppFEV1):_____
b. Body mass index (BMI):_____
c. Pulmonary exacerbations- number in preceding 6 months:_____

<u>Reauthorization</u>: 12 months with evidence of appropriate clinical response to therapy

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

a. Name of CF treating/consulting specialist/pulmonologist
_____
b. For consults, provide chart documentation including name of drug
_____

AND

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

a. For age appropriate patients, improved or stable lung function as demonstrated by percent predicted expiratory volume in 1 second (ppFEV1):_____
b. 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)