BlueCross BlueShield Federal Employee Program.

KALYDECO
(ivacaftor)

Pre - PA Allowance

None

Prior-Approval Requirements

Age 1 month of age or older

Diagnosis

The patient must have the following:

Cystic fibrosis (CF)

AND ALL the following:

1. Patient has one mutation in the *CFTR* gene that is responsive to Kalydeco (see Appendix 2)
2. **NO** homozygous for *F508del* mutation in the *CFTR* gene
3. Patients 6 years of age or older **only**: Pretreatment percent predicted forced expiratory volume (ppFEV1) must be provided
4. Baseline ALT and AST levels will be obtained and prescriber agrees to monitor every 3 months during the first year of treatment and annually thereafter
5. Must be prescribed by a pulmonologist or gastroenterologist
6. **NO** dual therapy with another cystic fibrosis transmembrane conductance regulator (CFTR) potentiator (see Appendix 1)

Prior - Approval Limits

Quantity 168 units per 84 days
Duration 12 months

Prior – Approval *Renewal* Requirements

Age 1 month of age or older

Diagnosis

Patient must have the following:

Cystic fibrosis (CF)

AND ALL of the following:

1. Patients less than 6 years of age **only**: Patient's symptoms have improved or stabilized from baseline
2. Patients 6 years of age or older **only**: Stable or improvement of ppFEV$_1$ from baseline
3. Prescriber agrees to monitor ALT and AST levels annually
4. **NO** dual therapy with another cystic fibrosis transmembrane conductance regulator (CFTR) potentiator (see Appendix 1)

Prior – Approval *Renewal* Limits

Same as above

Appendix 1 - List of Cystic Fibrosis Transmembrane Conductance Regulator (CFTR) Potentiators

Generic Name	Brand Name
ivacaftor	Kalydeco
ivacaftor/lumacaftor	Orkambi
ivacaftor/tezacaftor	Symdeko
ivacaftor/tezacaftor/elexacaftor	Trikafta

Appendix 2 - List of *CFTR* Gene Mutations that 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.
† 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.