BlueCross BlueShield Federal Employee Program

SYMDEKO
(tezacaftor and ivacaftor)

Pre - PA Allowance

None

Prior-Approval Requirements

Age 6 years of age or older

Diagnosis

Patient must have the following:

Cystic fibrosis (CF)

AND ALL of the following

1. Homozygous for the F508del mutation or at least one mutation in the CFTR gene that is responsive to Symdeko (see Appendix 2)
2. Pretreatment percent predicted forced expiratory volume (ppFEV$_1$) must be provided
3. Baseline ALT, AST, and bilirubin must be obtained at baseline and tested every 3 months for the first year
4. Must be prescribed by a pulmonologist or gastroenterologist
5. **NO** dual therapy with another cystic fibrosis transmembrane conductance regulator (CFTR) potentiator (see Appendix 1)

Prior - Approval Limits

Quantity 168 tablets for 84 days

Duration 6 months

Prior – Approval *Renewal* Requirements

Age 6 years of age or older

Diagnosis

Patient must have the following:

Cystic Fibrosis (CF)

AND ALL of the following:

1. Stable or improvement of ppFEV$_1$ from baseline
2. Annual testing of ALT, AST, and bilirubin levels after the first year of therapy
3. **NO** dual therapy with another cystic fibrosis transmembrane conductance regulator (CFTR) potentiator (see Appendix 1)

Prior - Approval *Renewal* Limits

Quantity 168 tablets for 84 days

Duration 12 months

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 Symdeko

546insCTA	*E92K*	*G576A*	*L346P*	*R117G*	*S589N*
711+3A→G	*E116K*	*G576A;R668C* †	*L967S*	*R117H*	*S737F*
2789+5G→A	*E193K*	*G622D*	*L997F*	*R117L*	*S912L*
3272-26A→G	*E403D*	*G970D*	*L1324P*	*R117P*	*S945L*
3849+10kbC→T	*E588V*	*G1069R*	*L1335P*	*R170H*	*S977F*
A120T	*E822K*	*G1244E*	*L1480P*	*R258G*	*S1159F*
A234D	*E831X*	*G1249R*	*M152V*	*R334L*	*S1159P*
A349V	*F191V*	*G1349D*	*M265R*	*R334Q*	*S1251N*
A455E	*F311del*	*H939R*	*M952I*	*R347H*	*S1255P*
A554E	*F311L*	*H1054D*	*M952T*	*R347L*	*T338I*

A1006E	*F508C*	*H1375P*	*P5L*	*R347P*	*T1036N*
A1067T	*F508C;S1251N* †	*I148T*	*P67L*	*R352Q*	*T1053I*
D110E	*F508del* ^	*I175V*	*P205S*	*R352W*	*V201M*
D110H	*F575Y*	*I336K*	*Q98R*	*R553Q*	*V232D*
D192G	*F1016S*	*I601F*	*Q237E*	*R668C*	*V562I*
D443Y	*F1052V*	*I618T*	*Q237H*	*R751L*	*V754M*
D443Y;G576A;R668C †	*F1074L*	*I807M*	*Q359R*	*R792G*	*V1153E*
D579G	*F1099L*	*I980K*	*Q1291R*	*R933G*	*V1240G*
D614G	*G126D*	*I1027T*	*R31L*	*R1066H*	*V1293G*
D836Y	*G178E*	*I1139V*	*R74Q*	*R1070Q*	*W1282R*
D924N	*G178R*	*I1269N*	*R74W*	*R1070W*	*Y109N*
D979V	*G194R*	*I1366N*	*R74W;D1270N* †	*R1162L*	*Y161S*
D1152H	*G194V*	*K1060T*	*R74W;V201M* †	*R1283M*	*Y1014C*
D1270N	*G314E*	*L15P*	*R74W;V201M;D1270N* †	*R1283S*	*Y1032C*
E56K	*G551D*	*L206W*	*R75Q*	*S549N*	
E60K	*G551S*	*L320V*	*R117C*	*S549R*	

^ A patient must have two copies of the *F508del* mutation or at least one copy of a responsive mutation presented above to be indicated.
† 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.