BlueCross BlueShield Federal Employee Program

ORKAMBI
(lumacaftor / ivacaftor)

Pre - PA Allowance

None

Prior-Approval Requirements

Age 1 year of age or older

Diagnosis

Patient must have the following:

Cystic Fibrosis (CF)

AND ALL of the following:

1. Homozygous for *F508del* mutation in the cystic fibrosis transmembrane regulator (CFTR) gene confirmed by FDA approved CF mutation test
2. Patients 6 years of age or older **only**: pretreatment percent predicted forced expiratory volume (ppFEV1) must be provided
3. Patients 6 years of age or older **only:** inadequate treatment response, intolerance, or contraindication to Symdeko (tezacaftor/ivacaftor)
4. Baseline levels of ALT, AST, and bilirubin must not be greater than 3x the upper limit of normal
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 336 tablets per 84 days **OR**
168 packets per 84 days

Duration 6 months

Prior – Approval *Renewal* Requirements

Age 1 year 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. Annual testing of ALT, AST, and bilirubin levels after the first year of therapy
4. **NO** dual therapy with another cystic fibrosis transmembrane conductance regulator (CFTR) potentiator (see Appendix 1)

Prior - Approval *Renewal* Limits

Quantity 336 tablets per 84 days **OR**
168 packets per 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