Commercial/Healthcare Exchange PA Criteria

Effective: November 22, 2019

Prior Authorization: Trikafta

Products Affected: Trikafta (elexacaftor/tezacaftor/ivacaftor) oral tablet therapy pack

Medication Description: Elexacaftor; tezacaftor; ivacaftor increases the quantity and function of the *F508del*-cystic fibrosis transmembrane conductance regulator (CFTR), a chloride channel present at the surface of epithelial cells in multiple organs, resulting in increases in chloride transport. Patients with cystic fibrosis (CF) have a mutation in the CFTR protein gene that encodes the protein. In patients with the F508del mutation, CFTR protein misfolding causes a defect in cellular processing and trafficking that targets the protein for degradation, resulting in a lower quantity of CFTR at the cell surface. The small amount of F508del-CFTR that does reach the cell surface is less stable and has low channel-open probability compared to the wild-type CFTR protein. Elexacaftor and tezacaftor facilitate the cellular processing and trafficking of normal and select mutant forms of CFTR (including F508del-CFTR) to increase the amount of mature CFTR protein delivered to the cell surface. Ivacaftor is a CFTR potentiator that increases chloride transport by potentiating the channel-opening probability of the CFTR protein. CFTR protein must be present at the cell surface for ivacaftor to function. Ivacaftor can potentiate the CFTR protein delivered to the cell surface by tezacaftor, leading to a further enhancement of chloride transport than either agent alone.

Covered Uses: Treatment of cystic fibrosis (CF) in patients aged 12 years and older who have at least one *F508del* mutation in the cystic fibrosis transmembrane conductance regulator *(CFTR)* gene.

Exclusion Criteria:

1. Concurrent therapy with Orkambi, Kalydeco, or Symdeko

Required Medical Information:

1. Diagnosis
2. Current therapy regimen
3. Cystic Fibrosis Transmembrane Regulator (CFTR) gene mutation (documentation required)

*If the patient's genotype is unknown, an FDA-cleared CF mutation test should be used to confirm the presence of at least one F508del mutation.

Age Restrictions: 12 years of age and older

Prescriber Restrictions: Prescribed by, or in consultation with, a pulmonologist or a physician who specializes in the treatment of Cystic fibrosis.

Coverage Duration: 3 years

Other Criteria:

A. Patient has a diagnosis of cystic fibrosis; AND
B. Patient has at least one F508del mutation in the cystic fibrosis transmembrane conductance regulator (CFTR) gene.

References:

1. Product Information: TRIKAFTA(TM) oral tablets, elexacaftor, tezacaftor, ivacaftor oral tablets; ivacaftor oral tablets. Vertex Pharmaceuticals Incorporated (per manufacturer), Boston, MA, 2019.

Policy Revision history

Rev #	Type of Change	Summary of Change	Sections Affected	Date
1	New Policy	New Policy	All	11/19/2019