

## SPECIALTY GUIDELINE MANAGEMENT

### TRIKAFTA (elexacaftor/tezacaftor/ivacaftor)

#### POLICY

##### I. INDICATIONS

The indications below including FDA-approved indications and compendial uses are considered a covered benefit provided that all the approval criteria are met and the member has no exclusions to the prescribed therapy.

###### FDA-Approved Indication

Trikafta is indicated for the treatment of patients with cystic fibrosis (CF) aged 2 years and older who have at least one F508del mutation in the cystic fibrosis transmembrane conductance regulator (*CFTR*) gene or a mutation in the *CFTR* gene that is responsive based on in vitro data.

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 or a mutation that is responsive based on in vitro data.

All other indications are considered experimental/investigational and not medically necessary.

##### II. REQUIRED DOCUMENTATION

The following information is necessary to initiate the prior authorization review: genetic testing report confirming the presence of the appropriate *CFTR* gene mutation.

##### III. CRITERIA FOR INITIAL APPROVAL

###### **Cystic Fibrosis**

Authorization of 6 months may be granted for treatment of cystic fibrosis when all of the following criteria are met:

- A. Genetic testing was conducted to detect a mutation in the *CFTR* gene.
- B. The medication is prescribed by or consultation with a pulmonologist.
- C. The member has one of the following mutations in the *CFTR* gene: A46D, A120T, A234D, A349V, A455E, A554E, A1006E, A1067T, D110E, D110H, D192G, D443Y, D443Y;G576A;R668C, D579G, D614G, D836Y, D924N, D979V, D1152H, D1270N, E56K, E60K, E92K, E116K, E193K, E403D, E474K, E588V, E822K, F191V, F311del, F311L, F508C, F508C;S1251N, F508del, F575Y, F1016S, F1052V, F1074L, F1099L, G27R, G85E, G126D, G178E, G178R, G194R, G194V, G314E, G463V, G480C, G551D, G551S, G576A, G576A;R668C, G622D, G628R, G970D, G1061R, G1069R, G1244E, G1249R, G1349D, H139R, H199Y, H939R, H1054D, H1085P, H1085R, H1375P, I148T, I175V, I336K, I502T, I601F, I618T, I807M, I980K, I1027T, I1139V, I1269N, I1366N, K1060T, L15P, L165S, L206W, L320V, L346P, L453S, L967S, L997F, L1077P, L1324P, L1335P, L1480P, M152V, M265R, M952I, M952T, M1101K, P5L, P67L, P205S, P574H, Q98R, Q237E, Q237H, Q359R, Q1291R, R31L, R74Q, R74W, R74W;D1270N, R74W;V201M, R74W;V201M;D1270N, R75Q, R117C, R117G, R117H, R117L, R117P, R170H, R258G, R334L, R334Q,

R347H, R347L, R347P, R352Q, R352W, R553Q, R668C, R751L, R792G, R933G, R1066H, R1070Q, R1070W, R1162L, R1283M, R1283S, S13F, S341P, S364P, S492F, S549N, S549R, S589N, S737F, S912L, S945L, S977F, S1159F, S1159P, S1251N, S1255P, T338I, T1036N, T1053I, V201M, V232D, V456A, V456F, V562I, V754M, V1153E, V1240G, V1293G, W361R, W1098C, W1282R, Y109N, Y161D, Y161S, Y563N, Y1014C, Y1032C, 3141del9, 546insCTA.

D. The member is at least 2 years of age.

E. Trikafta will not be used in combination with other ivacaftor containing medications.

#### **IV. CONTINUATION OF THERAPY**

Authorization of 12 months may be granted for continued treatment in members requesting reauthorization for an indication listed in Section III who are experiencing benefit from therapy as evidenced by disease stability or disease improvement (e.g., improvement in FEV1 from baseline).

#### **V. QUANTITY LIMIT**

1. Trikafta tablets have a quantity limit of 3 tablets per day.
2. Trikafta paks have a quantity limit of 2 packets per day.

#### **VI. REFERENCES**

1. Trikafta [package insert]. Boston, MA: Vertex Pharmaceuticals Inc; April 2023.