Effective Date: 4/2020
Reviewed: 01/2020, 2/2021, 7/2021, 2/2022, 2/2023, 7/2023, 2/2024, 5/2025
Scope: Medicaid

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. 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. Documentation that 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, 1507_1515del9, 2183A→G, A1067P, A107G, A309D, A62P,

C491R, D1445N, D565G, D993Y, E116Q, E292K, F1107L, F200I, F587I, G1047R, G1123R, G1247R, G27E, G424S, G480S, G551A, G970S, H620P, H620Q, H939R;H949L, I105N, I125T, I148N, I331N, I506L, I556V, K162E, K464E, L1011S, L137P, L333F, L333H, L441P, L619S, M1137V, M150K, N1088D, N1303I, N186K, N187K, N418S, P140S, P499A, P750L, Q1313K, Q372H, Q493R, Q552P, R1048G, R117C;G576A;R668C, R297Q, R31C, R516S, R555G, R709Q, R75L, S1045Y, S108F, S1118F, S1235R, S549I, T1086I, T1246I, T1299I, T351I, V392G, V603F, Y301C, 2789+5G→A, 3272-26A→G, 3849+10kbC→T, N1303K, 711+3A→G, E831X, 5T;TG12, 5T;TG13, 296+28A→G, 621+3A→G, 1898+3A→G, 2789+2insA, 3850-3T→G, 3600G→A, 3849+4A→G, 3849+40A→G, 4005+2T→C, 1341G→A, 3041-15T→G, 2752-26A→G .

D. The member is at least 2 years of age.
E. Trikafta will not be used in combination with other ivacaftor containing medications.

III. 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).

IV. 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.

Recommended Dosage for Adult and Pediatric Patients Aged 2 Years and Older (with fat-containing food (2.2, 12.3))			
Age	**Weight**	**Morning Dose**	**Evening Dose**
2 to less than 6 years	Less than 14 kg	One packet containing elexacaftor 80 mg/tezacaftor 40 mg/ivacaftor 60 mg oral granules	One packet containing ivacaftor 59.5 mg oral granules
	14 kg or more	One packet containing elexacaftor 100 mg/tezacaftor 50 mg/ivacaftor 75 mg oral granules	One packet containing ivacaftor 75 mg oral granules
6 to less than 12 years	Less than 30 kg	Two tablets, each containing elexacaftor 50 mg/tezacaftor 25 mg/ivacaftor 37.5 mg	One tablet of ivacaftor 75 mg
	30 kg or more	Two tablets, each containing elexacaftor 100 mg/tezacaftor 50 mg/ivacaftor 75 mg	One tablet of ivacaftor 150 mg
12 years and older	-	Two tablets, each containing elexacaftor 100 mg/tezacaftor 50 mg/ivacaftor 75 mg	One tablet of ivacaftor 150 mg

V. REFERENCES

1. Trikafta [package insert]. Boston, MA: Vertex Pharmaceuticals Inc; January 2025.