HAP CareSource™

PHARMACY POLICY STATEMENT
HAP CareSource™ Marketplace

DRUG NAME	Trikafta (elexacaftor, tezacaftor and ivacaftor; ivacaftor)
BENEFIT TYPE	Pharmacy
STATUS	Prior Authorization Required

Trikafta (elexacaftor, tezacaftor and ivacaftor; ivacaftor) is a combination of ivacaftor, a cystic fibrosis transmembrane conductance regulator (CFTR) potentiator, tezacaftor, and elexacaftor indicated for the treatment of cystic fibrosis (CF) in patients aged 2 years and older who have at least one F508del mutation in the CFTR gene or a mutation in the CFTR gene that is responsive based on in vitro data. The combined effect of elexacaftor, tezacaftor and ivacaftor is increased quantity and function of CFTR at the cell surface, resulting in increased CFTR activity as measured by CFTR mediated chloride transport.
Cystic fibrosis is an autosomal recessive disease in which patients can have abnormal airways secretions, chronic endobronchial infection, and progressive airway obstruction.

Trikafta (elexacaftor, tezacaftor and ivacaftor; ivacaftor) will be considered for coverage when the following criteria are met:

Cystic Fibrosis

For **initial** authorization:

1. Member is at least 2 years of age; AND
2. Medication must be prescribed by or in consultation with a pulmonologist or an infectious disease specialist; AND
3. Member has a diagnosis of cystic fibrosis; AND
4. Member has documentation of genetic testing in chart notes with at least one F508del mutation in the CFTR gene; OR
5. Member has at least **ONE** of the following mutations in the CFTR gene: 3141del9, E822K, G1069R, L967S, R117L, S912L, 546insCTA, F191V, G1244E, L997F, R117P, S945L, A46D, F311del, G1249R, L1077P, R170H, S977F, A120T, F311L, G1349D, L1324P, R258G, S1159F, A234D, F508C, H139R, L1335P, R334L, S1159P, A349V, F508C;S1251N, H199Y, L1480P, R334Q, S1251N, A455E, F508del, H939R, M152V, R347H, S1255P, A554E, F575Y, H1054D, M265R, R347L, T338I, A1006E, F1016S, H1085P, M952I, R347P, T1036N, A1067T, F1052V, H1085R, M952T, R352Q, T1053I, D110E, F1074L, H1375P, M1101K, R352W, V201M, D110H, F1099L, I148T, P5L, R553Q, V232D, D192G, G27R, I175V, P67L, R668C, V456A, D443Y, G85E, I336K, P205S, R751L, V456F, D443Y;G576A;R668C, G126D, I502T, P574H, R792G, V562I, D579G, G178E, I601F, Q98R, R933G, V754M, D614G, G178R, I618T, Q237E, R1066H, V1153E, D836Y, G194R, I807M, Q237H, R1070Q, V1240G, D924N, G194V, I980K, Q359R, R1070W, V1293G, D979V, G314E, I1027T, Q1291R, R1162L, W361R, D1152H, G463V, I1139V, R31L, R1283M, W1098C, D1270N, G480C, I1269N, R74Q, R1283S, W1282R, E56K, G551D, I1366N, R74W, S13F, Y109N, E60K, G551S, K1060T,

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R74W;D1270N, S341P, Y161D, E92K, G576A, L15P, R74W;V201M, S364P, Y161S, E116K, G576A;R668C, L165S, R74W;V201M;D1270N, S492F, Y563N, E193K, G622D, L206W, R75Q, S549N, Y1014C, E403D, G628R, L320V, R117C, S549R, Y1032C, E474K, G970D, L346P, R117G, S589N, E588V, G1061R, L453S, R117H, S737F, 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; AND

6. Baseline liver function tests (LFTs) have been or will be completed; AND
7. Provider attests member does **NOT** have severe hepatic impairment (Child-Pugh Class C).
8. **Dosage allowed/Quantity limit:** Quantity limit: 84 tablet or packet carton for 28 days.

Recommended Dosage for Adult and Pediatric Patients Aged 2 Years and Older			
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

If all the above requirements are met, the medication will be approved for 6 months.

For **reauthorization**:

1. Chart notes must show improvement or stabilized signs and symptoms of disease such as any of the following:
   a) Improved FEV1 and/or other lung function tests;
   b) Improvement in sweat chloride;
   c) Decrease in pulmonary exacerbations;
   d) Decrease in pulmonary infections;

e) Increase in weight-gain;
f) Decrease in hospitalizations.

If all the above requirements are met, the medication will be approved for an additional 12 months.

HAP CareSource considers Trikafta (elexacaftor, tezacaftor and ivacaftor; ivacaftor) not medically necessary for the treatment of conditions that are not listed in this document. For any other indication, please refer to the Off-Label policy.

DATE	ACTION/DESCRIPTION
11/12/2019	New policy for Trikafta created.
12/31/2020	New approved FDA mutations included. Diagnosis of cystic fibrosis added to initial criteria.
08/09/2021	Changed lower age limit to 6 years.
04/27/2022	Policy transferred to new template. Added dose information for children aged 6 to 11.
05/19/2023	Lowered age limit to 2 years to align with FDA approval; removed compliance with initial criteria and adherence in claims history from reauthorization criteria; added references.
01/23/2025	Added reference; added additional approved gene mutations per package insert; added baseline LFTs and absence of severe hepatic impairment per new black box warning per package insert; increased initial approval length to 6 months.

References:

1. Trikafta [prescribing information]. Boston, MA: Vertex Pharmaceuticals Inc; 2024.
2. Ren CL, Morgan RL, Oermann C, et al. Cystic Fibrosis Foundation Pulmonary Guidelines. Use of Cystic Fibrosis Transmembrane Conductance Regulator Modulator Therapy in Patients with Cystic Fibrosis. *Ann Am Thorac Soc*. 2018;15(3):271-280. doi:10.1513/AnnalsATS.201707-539OT.
3. Farrell PM, White TB, Ren CL, et al. Diagnosis of Cystic Fibrosis: Consensus Guidelines from the Cystic Fibrosis Foundation [published correction appears in J Pediatr. 2017 May;184:243]. J Pediatr. 2017;181S:S4-S15.e1. doi:10.1016/j.jpeds.2016.09.064
4. Southern KW, Castellani C, Lammertyn E, et al. Standards of care for CFTR variant-specific therapy (including modulators) for people with cystic fibrosis. J Cyst Fibros. 2023;22(1):17-30. doi:10.1016/j.jcf.2022.10.002

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