

## PHARMACY POLICY STATEMENT

BILLING CODE	Must use valid NDC
BENEFIT TYPE	Pharmacy
STATUS	Prior Authorization Required

Orkambi is a combination of ivacaftor, a cystic fibrosis transmembrane conductance regulator (CFTR) potentiator, and lumacaftor originally approved by the FDA in 2015. It is indicated for the treatment of cystic fibrosis (CF) in patients aged 1 year and older who are homozygous for the F508del mutation in the CFTR gene. If the patient's genotype is unknown, an FDA-cleared CF mutation test should be used to detect the presence of the F508del mutation on both alleles of the CFTR gene.

Orkambi (lumacaftor/ivacaftor) will be considered for coverage when the following criteria are met:

For \_\_\_\_\_:

Chart notes that show signs and symptoms of improvement, with any of the following:

- Improved FEV1 and/or other lung function tests;
- Improvement in sweat chloride;
- Decrease in pulmonary exacerbations;
- Decrease in pulmonary infections;
- Increase in weight-gain;
- Decrease in hospitalizations.

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

	New policy for Orkambi created. Not covered diagnosis added.
	Age coverage expanded (approved for 2 years old members and older).
	Diagnosis of cystic fibrosis added to initial criteria. Reauthorization criteria updated to ask for evidence of disease improvement. Added trial of Symdeko for members 6 years and older.
	Policy transferred to new template. Removed trial of Symdeko. Clarified dosing. Amended references.
	Age requirement expanded to 1 year of age and older. Updated dosing requirement.

References:

1. Orkambi [package insert]. Boston, MA: Vertex Pharmaceuticals Inc; September 2022.  
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. National Guideline Clearinghouse (NGC). Guideline summary: Cystic fibrosis pulmonary guidelines. Chronic medications for maintenance of lung health. In: National Guideline Clearinghouse (NGC) [Web site]. Rockville (MD): Agency for Healthcare Research and Quality (AHRQ); 2013 Apr 01. Available: <https://www.guideline.gov>.

Effective date: 04/01/2023

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