AETNA BETTER HEALTH®
Coverage Policy/Guideline

Name:	Orkambi	Page:	1 of 2
Effective Date:	3/24/2023	Last Review Date:	1/2023

Applies to:	☒Illinois	☐Florida	☒Florida Kids
	☐New Jersey	☒Maryland	☐Michigan
	☒Pennsylvania Kids	☒Virginia	☐Texas

Intent:

The intent of this policy/guideline is to provide information to the prescribing practitioner outlining the coverage criteria for Orkambi under the patient's prescription drug benefit.

Description:

Orkambi is a combination of lumacaftor and ivacaftor indicated for the treatment of cystic fibrosis (CF) in patients aged 1 year and older who are homozygous for the *F508del* mutation in the cystic fibrosis transmembrane conductance regulator (*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.

Limitation of use: The efficacy and safety of Orkambi have not been established in patients with CF other than those homozygous for the *F508del* mutation.

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

Applicable Drug List:

Orkambi

Policy/Guideline:

I. Submission of the following information is necessary to initiate the prior authorization review:

A. genetic testing report confirming the presence of the appropriate *CFTR* gene mutation.

Criteria for Initial Approval

Cystic Fibrosis

II. Authorization 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 member is positive for the F508del mutation on both alleles of the CFTR gene.
C. The member is at least 1 year of age.
D. Orkambi will not be used in combination with other medications containing ivacaftor.

Criteria for Continuation of Therapy

III. Reauthorization may be granted for with cystic fibrosis when the following has been met:

A. Member is experiencing benefit from therapy as evidenced by disease stability or disease improvement (e.g., improvement in FEV1 from baseline).

Approval Duration and Quantity Restrictions:

Approval: 12 months

Quantity Level Limit:

100 mg/125 mg: 112 tablets per 28 days

200 mg/125 mg: 112 tablets per 28 days

75 mg/94 mg granule packets: 56 packets per 28 days

100 mg/125 mg oral granule packets: 56 packets per 28 days

150 mg/188 mg oral granule packets: 56 packets per 28 days

References:

1. Orkambi [package insert]. Boston, MA: Vertex Pharmaceuticals Inc.; September 2022.